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OXFORD HANDBOOK OF GENERAL PRACTICE

Chantal Simon | Hazel Everitt
Francoise van Dorp | Nazia Hussain
Emma Nash | Danielle Peet

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General Practice

FIFTH EDITION

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Preface

For this new, fifth edition of the *Oxford Handbook of General Practice* (*OHGP*), we welcome three new guest editors:

Dr Nazia Hussain is a GP in Cardiff and a former Deputy Editor of the RCGP's educational journal for GPs in training, *InnovAiT*. She has revised the respiratory chapter for this edition of the *OHGP*.

Dr Emma Nash is a GP in Portchester. She is her CCG Mental Health Lead, a GP Training Programme Director, and one of the Clinical Leads working for the On-line Learning Environment team at the RCGP. She has revised the mental health chapter of this new edition of the *OHGP*.

Dr Danielle Peet is a GP in Manchester and was also a former Deputy Editor of *InnovAiT*. She has revised and separated out the chapters on cancer care and palliative care for this new edition of the *OHGP*.

Since the last edition of the *OHGP*, new areas of work are evolving for GPs. The concept of 'personalized' medicine driven by a deeper understanding of the genetic code underpinning our personal characteristics is a good example. The way that we work has also changed with a workforce crisis and many GPs now working outside traditional practice structures 'at scale'. Our teams have changed too, with practitioners undertaking new, extended-scope roles performing many of the tasks traditionally done by GPs; GPs are learning to manage much larger multidisciplinary teams, cope with increased complexity in their own consultations, and perform wider supervisory roles. Alongside these changes there has been a technology revolution, meaning that GPs now consult and communicate in a range of new ways.

To address all these factors, we have completely rewritten the non-clinical sections of the OHGP. We have simplified the structure to make information easier to find, and reduced the amount of detail on contracts as these are changing apace. We have also included new sections on new modes of consultation and communication. The clinical topics have all been updated and, in addition, we have included new sections on genetics and genomics, liver disease, multimorbidity, sepsis, risk scoring for GP emergencies, and communication across settings.

As always, we welcome feedback from our readers. We would also like to thank the many of you who have contacted us to point out errors, omissions, and ways to improve the *OHGP* in the past. It is thanks to you and the feedback that you provide that the *OHGP* continues to develop to meet your day-to-day needs.

CS
HE
FvD

Acknowledgements

This book would not have come into being without input from a large number of individuals.

First, we would like to thank our extremely helpful editorial team at Oxford University Press. In particular, we would like to thank Kate Smith, who has been a real pleasure to work with.

Next, we would like to thank our long-suffering families. Writing and editing a book like the *OHGP* is very time-consuming, and we all have other 'day jobs'. Without the support of our families, a project like the *OHGP* would not be possible. In alphabetical order, we would like to give special thanks to our close family: Ben Jewell, David Gough, Ian Wright, Jonathan Nash, Dr and Mrs M A Hussain, and Peter Wynn—and children: Adam, Alicia, Ben, Charlie, Chloe, Emily, Emma, Ethan, Hannah, Helena, Kate, Oskar, Samuel, and Sophie.

The *OHGP* has always had a 'hands-on' feel. That is because it is written by practising GPs who learn new things every day from their patients and colleagues. We would like to thank all those that we work with for their help and support. In particular, we would like to thank:

- The CPD, EKU/EKC, CIRC, and OLE teams at the RCGP
- The Department of Primary Care at the University of Southampton
- The Lansdowne Campus Library, GP Centre, and Health and Social Sciences Faculty of the University of Bournemouth
- The GPs, practice manager, staff, and patients of the Banks and Bearwood Medical Practice in Bournemouth
- The staff and patients of Westlands Medical Centre in Portchester

Finally, we would like to give special thanks to all the other many individuals who contributed, either directly or indirectly, to this edition of the *OHGP*. We do not always know who you are, as much of the *OHGP* is peer reviewed anonymously, but your expert input has been extremely valuable. In particular, we would like to thank:

- **The Primary Care Dermatology Society** for their hard work in helping us to substantially revise the Dermatology chapter of this book (Chapter 16). In particular: Dr Tim Cunliffe, Dr Helen Frow, Dr George Moncrieff, Dr Julian Peace, Dr Angelika Razzaque, Dr Chin Whybrew, Dr Iain Henderson and Dr Michelle Ralph
- **Mr Richard Newsom** (Independent Consultant Ophthalmologist) for his expert review of Chapter 25 (Ophthalmology)
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- **Dr Carmel Wills** (GP, Salisbury, Wiltshire) for her comprehensive feedback on the Emergencies chapter of this book (Chapter 29)

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- **Previous authors and editors of the *OHGP*** Dr Brian Stevenson, Dr Jon Birtwistle, Dr Knut Schroeder, Dr Matt Burkes, and Professor Tony Kendrick. **Our junior reviewing team** Dr Hossam Abdel-Hamid, Dr Hannah Billett, Dr Rosie Dudson, Dr Joshua Getty, Dr Abi Moore, Dr Smitha Thurairatnam, Dr Ravi Tomar, and Dr Adam Zacks.

If there are any omissions from this list, we apologize. Please tell us, and we will add your name to the list at the first opportunity.

CS

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Abbreviations used in the text

Evidence-based superscripts

N	NICE guideline
G	Guideline from a major guideline-producing body
C	Cochrane review
S	Systematic review or meta-analysis published in a major peer-reviewed journal
R	Randomized controlled trial published in a major peer-reviewed journal

Referral times

E	Emergency admission
U	Urgent referral
S	Soon referral
R	Routine referral

Handbook symbols

❗	Note
⚠	Warning
➲	*OHGP* cross reference
🖱	Web link
☎	Telephone number
♀	Female
♂	Male
💣	Controversy
▶▶	Don't dawdle
1°	Primary
2°	Secondary
↑	Increased/increasing
↓	Decreased/decreasing
→	Leading to/resulting in
~	Approximately
≈	Approximately equal
±	With or without

Standard abbreviations

AAA	abdominal aortic aneurysm
ABPI	ankle–brachial pressure index
ABPM	ambulatory blood pressure monitoring
ACE	angiotensin-converting enzyme
ACS	acute coronary syndrome
ACTH	adrenocorticotrophic hormone
ADH	antidiuretic hormone
ADHD	attention deficit hyperactivity disorder
AED	automated external defibrillator
AF	atrial fibrillation
AFP	alpha fetoprotein
AIDS	acquired immune deficiency syndrome
Alk phos	alkaline phosphatase
ALT	alanine aminotransferase
ANF	antinuclear factor
APH	antepartum haemorrhage
APMS	Alternative Provider Medical Services
ARB	angiotensin receptor blocker
ASD	atrial septal defect
ASO	antistreptolysin O
AST	aspartate aminotransferase
AV	arteriovenous
AXR	abdominal X-ray
BASHH	British Association for Sexual Health and HIV
BCG	Bacille Calmette–Guérin
bd	twice daily
BMA	British Medical Association
BMJ	*British Medical Journal*
BNF	*British National Formulary*
BP	blood pressure
bpm	beats per minute
Ca^{2+}	calcium
CABG	coronary artery bypass graft
CCF	congestive cardiac failure
CCG	Clinical Commissioning Group
CF	cystic fibrosis
CHC	combined hormonal contraception

CHD	coronary heart disease
CIN	cervical intraepithelial neoplasia
CMV	cytomegalovirus
CNS	central nervous system
COC	combined oral contraceptive
COPD	chronic obstructive airways disease
Cr	creatinine
CRP	C-reactive protein
CT	computed tomography
CVA	cerebrovascular accident
CVD	cardiovascular disease
CXR	chest X-ray
d	day(s)
DDH	developmental dysplasia of the hip
DES	Directed Enhanced Service
DH	Department of Health and Social Care
DIPJ	distal interphalangeal joint
DLA	Disability Living Allowance
DI	diabetes insipidus
DM	diabetes mellitus
DN	district nurse
DOAC	direct-acting oral anticoagulant
DRE	digital rectal examination
DVLA	Driving and Vehicle Licensing Authority
DVT	deep vein thrombosis
EBV	Epstein–Barr virus
ECG	electrocardiogram
Echo	echocardiogram
EEG	electroencephalogram
ENT	ear, nose, and throat
EPAU	early pregnancy assessment unit
ESR	erythrocyte sedimentation rate
ESRD	end-stage renal disease
FBC	full blood count
FBG	fasting blood glucose
FEV_1	forced expiratory volume in 1 second
FH	family history
FSH	follicle-stimulating hormone
FSRH	Faculty of Sexual and Reproductive Healthcare
FVC	forced vital capacity
g	grams

GA	general anaesthetic
GI	gastrointestinal
GGT	gamma glutamyl transferase
GMC	General Medical Council
GMS	General Medical Services
GP	general practitioner
GPC	General Practitioner Committee
GTN	glyceryl trinitrate
GTT	glucose tolerance test
GU	genitourinary
GUM	genitourinary medicine
h	hours
Hb	haemoglobin
HbA1c	glycosylated haemoglobin
HBPM	home blood pressure monitoring
HBsAg	hepatitis B surface antigen
hCG	human chorionic gonadotropin
HDL	high-density lipoprotein
Hib	*Haemophilus influenzae* type b
HIV	human immunodeficiency virus
HOCM	hypertrophic obstructive cardiomyopathy
HPV	human papilloma virus
HRT	hormone replacement therapy
HSV	herpes simplex virus
HV	health visitor
HVS	high vaginal swab
ICP	intracranial pressure
Ig	immunoglobulin
IHD	ischaemic heart disease
IM	intramuscular
INR	international normalized ratio
IT	information technology
IU	international units
IUCD	copper intrauterine device
IUD	intrauterine device
IUS	progestogen-containing intrauterine system
IV	intravenous
IVP	intravenous pyelogram
JVP	jugular venous pressure
K^+	potassium
kg	kilograms

KUB	kidney, ureters, and bladder X-ray
L	litres
LA	local anaesthetic
LBBB	left bundle branch block
LFT	liver function test
LH	luteinizing hormone
LIF	left iliac fossa
LMP	last menstrual period
LMWH	low-molecular-weight heparin
LN	lymph node
LRTI	lower respiratory tract infection
LTOT	long-term oxygen therapy
LUQ	left upper quadrant
LVF	left ventricular failure
LVH	left ventricular hypertrophy
m	metres
MAOI	monoamine oxidase inhibitor
M,C&S	microscopy, culture, and sensitivity
MCP	metacarpophalangeal
MCV	mean cell volume
MDI	metered dose inhaler
mg	milligrams
MHRA	Medicines and Healthcare products Regulatory Agency
MI	myocardial infarction
min	minutes
mL	millilitres
MMR	measles, mumps, and rubella
MND	motor neurone disease
mmHg	millimetres of mercury
mo	months
MRI	magnetic resonance imaging
MS	multiple sclerosis
MSU	midstream urine
Na^+	sodium
NAAT	nucleic acid amplification test
NHS	National Health Service
NI	National Insurance
NICE	National Institute for Health and Care Excellence
nocte	at night
NSAID	non-steroidal anti-inflammatory drug
NSTEMI	non-ST elevation myocardial infarction

O_2	oxygen
OA	osteoarthritis
OCD	obsessive–compulsive disorder
od	once daily
OOH	out-of-hours
OT	occupational therapy
OTC	over-the-counter
PAN	polyarteritis nodosa
PCI	percutaneous coronary intervention
PCO	primary care organization
PCOS	polycystic ovarian syndrome
PD	Parkinson’s disease
PE	pulmonary embolus
PEFR	peak expiratory flow rate
PET	pre-eclamptic toxaemia
PHCT	primary healthcare team
PIP	Personal Independence Payment
PIPJ	proximal interphalangeal joint
PMH	past medical history
PMS	Personal Medical Services
PN	practice nurse
po	oral
PO_4^{3-}	phosphate
POP	progesterone-only pill
PPE	personal protective equipment
PPH	post-partum haemorrhage
PR	per rectum
prn	as needed
qds	four times daily
QOF	Quality and Outcomes Framework
RA	rheumatoid arthritis
RBBB	right bundle branch block
RCGP	Royal College of General Practitioners
RCOG	Royal College of Obstetricians and Gynaecologists
RhD	rhesus factor
RIF	right iliac fossa
RPOCT	rapid point-of-care test
RR	relative risk
RTA	road traffic accident
RUQ	right upper quadrant
s	seconds

sc	subcutaneous
SH	sexual health
SLE	systemic lupus erythematosus
SOL	space-occupying lesion
SNRI	serotonin and noradrenaline reuptake inhibitor
SSRI	selective serotonin reuptake inhibitor
stat	immediately
SpO_2	peripheral oxygen saturation
STEMI	ST elevation myocardial infarction
STI	sexually transmitted infection
SVC	superior vena cava
TB	tuberculosis
TCA	tricyclic antidepressant
tds	three times daily
TFT	thyroid function tests
TIA	transient ischaemic attack
u	units
U&E	urea and electrolytes
UC	ulcerative colitis
URTI	upper respiratory tract infection
US(S)	ultrasound (scan)
UTI	urinary tract infection
VF	ventricular fibrillation
VSD	ventriculoseptal defect
VT	ventricular tachycardia
WCC	white cell count
wk	weeks
y	years

❗ *All other abbreviations are defined in the text on the page in which they appear.*

Elderly care and child health flags: conditions peculiar to children or elderly people, or in which management varies for these groups, are flagged as follows:

Child health

Elderly care

Chapter 1

What is general practice?

What is general practice? *2*
General practice in the UK *4*
Becoming a GP in the UK *6*
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What is general practice?

'Generalism describes a philosophy of practice which is person, not disease, centred; continuous, not episodic; integrates biotechnical and biographical perspectives; and views health as a resource for living and not an end in itself.'[1]

In the early 19th century, when apothecaries, physicians, and surgeons provided medical care, the term 'general practitioner' became applied to apothecaries taking the Membership Examination of the Royal College of Surgeons of England. Over the past 60y, general practice has established itself as the cornerstone of most national healthcare systems. In general, the better the primary care provision, the better the health of a population.[2] General practitioners (GPs or family physicians) have shown the intellectual framework within which they operate is different from, complementary to, but no less demanding than that of specialists.

What is medical generalism? The RCGP defines medical generalism as: 'An approach to the delivery of health care that routinely applies a broad and holistic perspective to the patient's problems.' It involves:

- Seeing the person as a whole and in the context of his/her family and wider social environment
- Using this perspective as part of the clinical method and therapeutic approach to all clinical encounters
- Being able to deal with undifferentiated illness and the widest range of patients and conditions
- In the context of general practice, taking continuity of responsibility for people's care across many disease episodes and over time and coordinating care across both health and social care organizations

The role of the GP GPs diagnose illness, treat minor illness in the community, promote better health, prevent disease, diagnose and certify disease, monitor chronic disease, and refer patients requiring specialist services. General practice is the primary access point to health services.

Although patients have an average of >5.5 consultations with their GP every year in the UK, only 1 in 20 consultations results in a secondary care referral. To do this, GPs must:

- Have a working knowledge of the whole breadth of medicine
- Maintain ongoing relationships with their patients—they are the only doctors to remain with their patients through sickness and health
- Focus on patients' response to illness rather than the illness itself taking into account personality, family patterns, and the effect of these on the presentation of symptoms
- Be interested in the ecology of health and illness within communities and in the cultural determinants of health beliefs
- Be able to draw on a wide range of resources including intuition, medical knowledge, communication skills, business skills, and humanity

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[1] Reeve J (2010) Protecting generalism—moving on from evidence-based medicine? *BJGP* **60**:521.

[2] Starfield B, Shi L, Macinko J (2005) Contribution of primary care to health systems and health. *Milbank Q* **83**:457–502.

In addition to day-to-day medical care of their patients, GPs in the UK have a number of additional roles:

- **Gatekeeping** GPs control access to hospital-based services enabling cost-effective care
- **Navigating** GPs work with patients/carers to guide them effectively and safely through the healthcare system
- **Service redesign and improvement** GPs manage service provision within their own practices, and beyond their practice boundaries
- **Research** GPs need critical appraisal skills to understand and apply relevant evidence to inform clinical decision-making. They need to be competent in collecting and analysing data for service improvement, and must collaborate effectively in primary care-based research
- **Education** GPs can be effective teachers in a wide range of contexts, educating patients, practice staff, medical students and junior doctors, fellow GPs, and the general public
- **Leadership** Many GPs have leadership roles—in their own practices, within their localities, or nationally

What is the difference between GPs and specialists? Marshall Marinker contrasted the role of generalists and specialists as follows:

GPs	Specialists
Exclude the presence of serious disease	Confirm the presence of serious disease
Tolerate uncertainty—managing patients with undifferentiated symptoms	Reduce uncertainty—investigating until a diagnosis is reached
Explore probability of seeing patients from a population with a relatively low incidence of serious disease	Explore possibility of seeing a pre-selected population of patients with a relatively high incidence of serious disease
Marginalize danger—recognizing and acting on danger signs even when diagnosis is not certain	Marginalize error—ensuring accurate diagnosis and treatment

To perform their roles well, GPs must show empathy for their patients; engagement and commitment to involve themselves in every aspect of patient care; appreciation of the limits of their skills and expertise; and professionalism in their dealings with both patients and colleagues.

What is primary care? Primary care services provide the first point of contact in a healthcare system. In the UK, general practice is one of the primary care services; others include community pharmacy, dental services, and optometry services.

Further information

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General practice in the UK

In 1948 the National Health Service (NHS) was formed, giving free healthcare for the entire population of the UK paid for by the taxpayer. Its structure varies from country to country within the UK. However, GPs form the 'front-line' of the NHS in all four nations, providing primary medical care and acting as 'navigators' to the rest of the health/social care system.

Workload ~97% of the British population is registered with a GP. Patients register with a practice of their choice—whole families are often registered with the same practice. Once registered, patients stay with that practice for an average of 12y. GPs carry out ~300 million consultations/y in England alone: 84% at the surgery, 12% by telephone, and 4% at the patient's home. 70% of the GP's total workload is spent with a patient, while >20% is currently spent on administration.

Working hours Standard practice working hours are 8 a.m.–6.30 p.m. on normal working weekdays. GP days are usually broken into 2 'sessions', each lasting a notional 4h 10min. A traditional working day for a GP involves 2 surgeries seeing patients with time for home visiting and administrative work between. However, the UK government has committed to extend routine primary care access into evenings and weekends and different models to achieve this are evolving. With these changes, the concept of 'sessions' and standard working hours is being lost.

Primary care provider or 'practice' Terms used to designate any organization providing NHS primary care services.

Practice list All patients registered with a particular primary care provider. Lists may be *open* (accepting new patients) or, by agreement with the PCO for a set period of time, *closed* to new patients—➲ p. 40.

Practice boundaries Traditionally, practices have set geographical boundaries agreed with their PCOs. The practice only accepts new patients onto the practice list who live within that boundary. Practices are also required to set 'outer boundaries' to enable existing patients who have moved home to stay with their practice.

Patient Choice Scheme Since 2015, practices in England may accept out-of-area registrations without obligation to provide home visiting services. If too ill to travel to their registered practice, patients can access urgent medical care via the NHS 111 service.

Primary care contracts The provider contract with the local PCO defines services primary care providers will provide, standards to achieve, and payment they will receive. Contract models: ➲ p. 32.

Commissioning Over the past 10y in the UK, mechanisms have evolved to enable local commissioning of services to meet local needs. Commissioning aims to design improved patient pathways while enabling more efficient use of funds. The commissioning cycle involves:

- Planning Establishing what local services exist and what is needed
- Commissioning services To match resources to need
- Monitoring Ensuring that service delivery meets expectations
- Revision Regular review of services to provide best possible care

Primary care/GP networks and federations Multiple practices coming together in some form of collaboration. Many different models are in operation from loose alliances to formation of companies. Working at scale enables sharing of costs and resources, greater flexibility, more protection against financial pressures, and the capacity to offer extended or new services for patients in the community.

Models of care in England The 'NHS Five-year Forward View' (2014) set out a blueprint for change in NHS England, setting out 7 new models of more integrated care including:

- Multispecialty community provider (MCP) GPs and practices working together in networks/federations and collaborating with other health and social care professionals from primary and secondary care to provide more integrated services in the community
- Urgent and emergency care networks Emergency departments in hospitals, working together with GP out-of-hours services, urgent care centres, minor injury units, NHS 111, and ambulance services to ensure people with urgent care needs get the right level of support in the right place at the right time, making best use of limited resources
- Enhanced health in care homes Brings together care home providers, NHS, and social care services to improve care for this vulnerable section of the community
- Primary and acute care system (PACS) A single provider or group of providers working together take responsibility for delivering the full range of primary, community, mental health, and hospital services for their local population

Sustainability and Transformation Partnership/Plan (STP) 44 covering England. Local NHS organizations and councils working together to draw up a plan to improve health and care in the areas they serve. The plan must set out practical ways to achieve better health and improved quality of care delivery within financial constraints.

Accountable care systems/organization (ACS) An evolved version of a Sustainability and Transformation Partnership in England. A single body—usually comprising NHS organizations ± local authorities—takes responsibility for the entire local healthcare system. This body holds the whole local budget and both commissions and provides services to meet the health (± social) care needs of its local population, taking complete responsibility for resources and population health.

Integrated care in Scotland, Wales, and Northern Ireland While the 'NHS Five-year Forward View' only applies to England, similar changes are occurring across the UK with a move to larger primary care networks of providers, providing a wider range of services over extended hours and offering integrated primary, secondary, and social care.

Further information

NHS England (2014) NHS Five Year Forward View. 🖱 https://www.england.nhs.uk/wp-content/uploads/2014/10/5yfv-web.pdf

NHS England (2017) Next Steps on the NHS Five Year Forward View. 🖱 http://napc.co.uk/wp-content/uploads/2017/09/NEXT-STEPS-ON-THE-NHS-FIVE-YEAR-FORWARD-VIEW.pdf

Becoming a GP in the UK

GP vocational training Involves 3y full-time (or equivalent part-time) specialty training after the Foundation years; 18mo in selected hospital specialty posts and 18mo as a *GP registrar* in general practice.

Recruitment and selection There are 2 rounds of GP recruitment each year in the UK. Applications are made online to the National Recruitment Office for General Practice Training (NRO):

- All applicants meeting GP training entry criteria are invited to sit a machine-markable test, the Multi-Specialty Recruitment Assessment (MSRA); candidates scoring ≥575 move directly to 'offer' stage
- Candidates scoring <575 on the MSRA attend a selection centre at their preferred location. Selections centres in England are the regional deaneries; Scotland, Wales, and Northern Ireland each have a single selection centre. Assessment comprises 3 simulation exercises and a written exercise. Following assessment, the highest ranked applicants are offered training places in that area. Appointable candidates without training places can opt to select other areas with remaining places

Criteria for training practices Each region sets its own criteria for selection of practices to train prospective GPs. Practices must have a suitably qualified GP trainer, and demonstrate high-quality administration, clinical care, and commitment/capability to educate a GP registrar. Details of local requirements can be obtained from regional deanery offices.

RCGP GP curriculum Consists of a single curriculum statement, 'Being a General Practitioner', covering the core capabilities and competences of a GP. This is supported by a series of 'topic guides':

Professional capabilities

- Consulting in general practice
- Equality, diversity, and inclusion
- Evidence-based practice, research, and sharing knowledge
- Improving quality, safety, and prescribing
- Leadership and management
- Urgent and unscheduled care

Stages of life

- Children and young people
- Reproductive health and maternity
- People living with long-term conditions (including cancer)
- Older adults
- People at the end of life

Clinical issues

- Alcohol and substance misuse
- Cardiovascular disease
- Dermatology
- ENT, oral, speech, and hearing
- Eyes and vision
- Gastroenterology
- Genetics/genomics
- Gynaecology and breast disease
- Haematology
- Immunology and allergy
- Infectious disease and travel health
- Mental health
- Metabolic problems and endocrinology
- Musculoskeletal
- Neurology
- Neurodevelopmental disorders, intellectual disability, and social disability
- Population health
- Renal medicine and urology
- Respiratory
- Sexual health

Membership of the RCGP (MRCGP) The MRCGP is an examination of professional competency based on the GP curriculum. Passing the 3 different components of the MRCGP is compulsory for all doctors wishing to become GPs in the UK:

Applied Knowledge Test (AKT) 200 multiple-choice questions test whether the candidate can apply knowledge in the context of general practice. Computer-based assessment held at Pearson VUE centres across the UK. Questions are distributed as follows: clinical medicine (80%); administration and informatics (10%); and research, appraisal, evidence-based medicine, and statistics (10%).

Clinical Skills Assessment (CSA) Mock surgery in which patients are played by actors. The candidate performs 13 consultations each of 10min duration while being observed by an examiner. Only available at RCGP Euston; takes place in February, May, and November each year.

Workplace-Based Assessment (WPBA) Continuous qualitative assessment of performance in training based on the Trainee ePortfolio (TeP) and trainer's report. Evidence collected is reviewed ≥1×/12mo by the deanery Annual Review of Competence Progression (ARCP) panel to ensure the trainee is ready to move to the next year of training.

Certification of Completion of Training (CCT) During training, on completion of each placement, the clinical supervisor completes an assessment of the trainee's performance. The GP educational supervisor signs this assessment off as confirmation that there has been satisfactory progress in acquiring the relevant curriculum competencies. The portfolio of assessments is reviewed and endorsed by the deanery at least annually (ARCP review). Submission of this portfolio together with successful completion of the MRCGP enables a GP in training to apply for the CCT via the RCGP certification unit.

RCGP Certification Unit Evaluates general practice training and makes recommendations for CCTs to the GMC. Anyone undertaking a training programme leading to a CCT should register with the Certification Unit.

Certification of Eligibility for GP Registration (CEGPR) An alternative route for doctors who have completed the CCT programme and passed either the AKT or CSA after leaving the programme, or are not eligible for a CCT but believe that their training, qualifications, and experience are equivalent (e.g. all or part of their training was outside the UK, or in a non-approved training post). Trainees who begin a 3y planned programme for a CCT and decide to shorten it, by including posts not in their GP programme, must also apply for a CEGPR. This type of application is more complex and time-consuming than the CCT route. Application is online to the GMC with submission of a portfolio of evidence.

Further information

National Recruitment Office for General Practice Training (NRO)
🖱 https://gprecruitment.hee.nhs.uk/
RCGP ☎ 020 3188 7400 🖱 www.rcgp.org.uk

Education in primary care

Education is vitally important in primary care to keep healthcare professionals up to date with both clinical and administrative/managerial parts of their roles, and to provide career development and direction.

Teaching in general practice Many GPs are involved in teaching: medical students, foundation doctors, GPs in training, new practice staff, and/or their peers. Teaching can be very rewarding but also brings stresses (e.g. preparation of material). Payments are available to GPs who take medical students, foundation doctors, and/or GPs in training into their surgeries for teaching and there are a few teaching posts within UK universities for GPs.

❗ As a teacher it is your responsibility to ensure you are competent to fulfil the task. Take steps to acquire proficiency in teaching skills. Local medical schools often run courses for prospective teachers.

Foundation doctors ➔ p. 20 **GP training** ➔ p. 6

Principles of self-directed and adult learning The learner takes responsibility for defining learning needs, setting goals, identifying resources, implementing appropriate activities, and evaluating outcomes. Adults are motivated by education that:

- Is based on mutual trust and respect
- Can be immediately applied in practice
- Involves cycles of action and reflection
- Allows them to take responsibility for their own learning
- Is based on, and builds on, previous experience
- Actively involves them
- Is perceived as relevant
- Is focused on problems

Learning style It has been proposed that understanding an individual's learning style helps tailor educational activities to be most effective—but this has been contested as we all apply different learning styles in different situations. Several learning style models are used and relate to personality traits. *Examples*:

Honey and Mumford's learning styles
- Activists Get involved, open-minded, enthusiastic
- Reflectors Stand back, think, cautious
- Theorists Look for principles, logical, perfectionist
- Pragmatists Practical, experimental, down to earth

Kolb's learning styles
- Divergers '*Why*' (concrete, reflective)—learn best when they know why something is relevant and how it will apply to their work
- Assimilators '*What*' (abstract, reflective)—learn best when given plenty of time to think and link different concepts in their minds
- Convergers '*How*' (abstract, active)—like to work actively on well-defined tasks and learn by trial and error
- Accommodators '*What if*' (concrete, active)—learn best by applying course material in new situations and solving problems they create for themselves

Continuing professional development (CPD) Aims to help GPs to provide high-quality patient care throughout their careers. Doctors need to demonstrate that they have up-to-date knowledge across the spectrum of general practice to become a registered GP and then need to show they are continuing to update and expand their knowledge to meet the requirements of appraisal and revalidation (➔ p. 10).

Personal development plans (PDPs) Outline areas of knowledge in need of update and ways those needs can be met. PDPs are an integral part of junior doctor training, GP training, and the appraisal process. Ask:

- *What* you need to learn—specific, measurable objectives (Box 1.1)
- *Why* you need to learn it and *how* you plan to learn it
- *How* you will know *whether* you have learnt it
- How your intentions link to *past* and *future* learning

Box 1.1 'SMART' criteria—learning objectives should be:

Specific
Measurable
Achievable
Realistic
Timed (i.e. there should be a deadline for achieving them)

Experiential learning—Figure 1.1. Learning through experience is a continuous process. It is the largest single source of learning for professionals throughout their working lives. To optimize learning, it is important to actively seek appropriate experiences, ask for feedback, relate the experience to your existing knowledge base, and reflect.

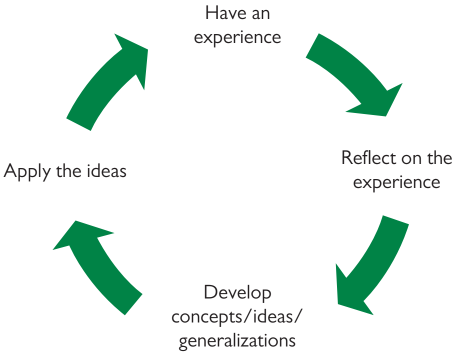
Have an experience
Reflect on the experience
Develop concepts/ideas/ generalizations
Apply the ideas

Figure 1.1 The Kolb cycle of learning

Reflection This is an inherent part of professionalism. It enriches experiential learning and challenges assumptions, enabling experience to be transformed into knowledge, skills, and attitudes. The Gibbs reflective cycle can be a useful tool to facilitate reflection:

- Description What happened?
- Feelings What do you think and feel about the event described?
- Evaluation What was good and bad about the experience?
- Analysis What sense can you make of this situation?
- Conclusion What did you do well? What else could you have done?
- Action plan What are you going to do as a result? If the same situation arose again, what would you do differently next time?

Appraisal and revalidation

Appraisal Requires all doctors wishing to practise medicine in the UK to undergo a formal review on a yearly basis. It is the basis of revalidation to maintain a licence to practise and aims to:

- Set out personal and professional development needs, career paths, and goals, and agree plans for them to be met
- Review the doctor's performance and consider the doctor's contribution to quality and improvement of local healthcare services
- Optimize the use of skills and resources in achieving the delivery of high-quality care
- Offer an opportunity for doctors to discuss and seek support for their participation in activities
- Identify the need for adequate resources to enable service objectives to be met

Supporting information The supporting information that doctors need to bring to the appraisal falls under 4 broad headings:

- General information—provides context on all aspects of work
- Keeping up to date—maintaining/enhancing quality of work
- Review of practice—evaluating quality of current practice
- Feedback on practice—how others perceive your work

Based on the GMC's document 'Good Medical Practice', there are six types of supporting information GPs are expected to provide and discuss at the appraisal at least once in each 5y cycle. They are:

- Continuing professional development
- Quality improvement activity
- Review of complaints/compliments
- Significant events
- Feedback from colleagues
- Feedback from patients

The Appraiser Responsible officers of designated bodies are accountable for ensuring appraisal takes place. Appraisers are properly trained to carry out this role and are in a position to undertake appraisal of a doctor's whole practice, including clinical performance, and where appropriate, specialist aspects of performance, e.g. research, service delivery, or management issues. Appraisers for GPs will generally be other GPs.

The appraisal process

- **Before the interview** Doctors must prepare an appraisal document containing information and supporting evidence about their practice and personal needs. Folders should be submitted to the appraiser ≥2wk prior to appraisal interviews to allow adequate time for preparation. Various electronic toolkits and portfolios are available
- **At the interview** Doctor and appraiser agree a summary of achievement in the past year, objectives for the next year, key elements of a personal development plan, and actions expected of the organization
- **After the interview** A summary document is produced and a joint declaration signed that the appraisal has been carried out properly

Licence to Practise In the UK, the GMC introduced licences to practise in November 2009. All registered doctors were able to request a licence to practise; all doctors eligible for registration with the GMC since November

2009 have also been licensed. The GMC licence (rather than GMC registration) signifies to patients that a doctor has the legal authority to write prescriptions and sign death certificates etc. GPs working in the NHS, either on a permanent or locum basis, need to be:

- Licensed by the GMC
- Listed on the GMC's General Practice Register
- Included on an NHS Performers List

Revalidation Revalidation was formally introduced in December 2012. All licensed doctors need to be relicensed every 5y.

Process of revalidation Doctors need to provide supporting information that shows that they keep up to date and remain fit to practise. GPs are accountable to their local 'designated body' and 'responsible officer'. In order for responsible officers to recommend maintenance of a GP's licence to practise, they need to be satisfied that:

- The GP has participated in an annual appraisal process that covers all of their medical practice
- There are no unresolved concerns about the doctor's performance

Responsible officer recommendations to the GMC

- **Positive recommendation** The doctor should be revalidated and his/her licence to practise continued
- **Deferral: insufficient information** Revalidation cannot be recommended because the doctor has not provided enough information; the doctor will be asked to provide additional information
- **Notification of failure to engage** The doctor has failed to engage with local systems and processes that support revalidation

The GMC will withdraw a licence to practise if

- The doctor tells them that it is no longer required
- The doctor does not pay the appropriate fee
- The doctor does not take part in the revalidation process when asked
- A Fitness to Practise Panel directs that the doctor's registration should be suspended or erased

❗ Doctors have a right of appeal against any decision to withdraw, or refuse to restore, their licence to practise or specialist certificate.

⚠ **Concerns about performance** Any GP with concerns about their own, or a colleague's, performance should discuss the matter confidentially with the secretary of their LMC, the clinical governance lead/performance information manager of their PCO, or the GMC.

Further information

GMC 🖱 Revalidation. www.gmc-uk.org/doctors/revalidation.asp

RCGP (2014) Principles of GP appraisal. 🖱 www.rcgp.org.uk/-/media/Files/Revalidation-and-CPD/CPD-Credits-and-Appraisal/The-Principles-of-GP-Appraisal-for-Revalidation-2014.ashx?la=en

RCGP (2016) Guide to supporting information for appraisal and revalidation. 🖱 www.rcgp.org.uk/-/media/Files/Revalidation-and-CPD/2016/RCGP-Guide-to-Supporting-Information-2016.ashx?la=en

GP working arrangements

Primary Care Performer List List of all doctors deemed competent to provide primary medical care held by the PCO.

Partnership Traditionally, GPs in the UK have worked as independent, self-employed contractors providing core primary healthcare services and additional services as negotiated within their contract.

Groups of self-employed contractors working together for mutual benefit are termed partnerships. A partnership can become a primary care provider as long as ≥1 partner is a GP. Although traditionally partnerships are made up of GPs only, practice managers, nurses, allied health professionals, and pharmacists can be included within partnerships.

Partners not only have responsibility to provide medical care, they also have management responsibilities for staff, premises, and equipment. Most receive a profit share, so the amount each partner is paid depends not only on income to the practice, but also expenditure:

Income

- Income from the NHS GMS, PMS, or APMS contract work
- Private work Includes private appointments (e.g. clinical assistant, industrial appointments); insurance examinations/reports; private medical examinations and certificates (e.g. HGV licence applications)

Expenditure

- Running costs of the practice Staff salaries; premises (rent, rates, repairs, maintenance, insurance); service costs (heating, water, electricity, gas and telephone bills, stationery and postage); training costs etc.
- Capital expenses Purchase of new medical and office equipment

Salaried GP A GP employed by a PCO, practice, or APMS. PCOs and GMS practices are bound by a nationally agreed model contract, with a salary within a range set by the Review Body. PMS practices and APMS providers can make their own arrangements. Salaried posts have advantages for those who do not want to take the financial risk or commit to long-term working within 1 practice.

Freelance GP or locum Provides medical cover on an ad hoc basis. Tend to be self-employed and may work independently, or through a locum chambers or agency. A charge is made for each session worked. Long-term locums should make their own pension provision or apply to join the NHS scheme.

GP with Extended Role (GPwER) A GP doing work that is:

- Beyond the scope of GP training and the MRCGP, and that a GP cannot carry out without further training, or
- Undertaken within a contract or setting that distinguishes it from standard general practice, or
- Offered for a fee outside of care to the registered practice population (teaching, training, research, occupational medicals, medico-legal reports, cosmetic procedures, etc.)

With expansion of care provision out of hospital, increasing numbers of GPs are taking up GPwER roles. A national accreditation scheme is currently being developed by the RCGP.

GP Registrar ➔ p. 6

Flexible career schemes In a bid to make general practice a better career option for doctors, in many areas of the UK flexible career options are being offered to both GPs just finishing their training and established GPs. Different schemes exist in different locations, but most combine part-time traditional general practice with another role, e.g. leadership, education, or a specialist clinical role.

GP Retention Scheme Provides opportunity for GPs who might otherwise leave the profession to remain working in general practice. GPs are eligible if:

- They hold full GMC registration, are on the Performers List, and want to change, or have changed, their hours due to caring responsibilities, because they are approaching retirement, or to enable them to take up other work within or outside general practice
- A regular part-time role does not meet their need for flexibility (e.g. need for short clinics or annualized hours)
- There is a need for additional educational supervision (e.g. pro rata study leave is inadequate to maintain professional competence)

Retained GPs work 1–4 sessions/wk (of 4h 10min) for up to 5y subject to annual review of eligibility; they receive an allowance of £1000–£4000/y depending on number of sessions worked plus usual pay. Employing practices receive a sum for each session worked (currently £76.92).

Further information

BMA GP Retention Scheme: step-by-step guide. 🖱 https://www.bma.org.uk/advice/employment/gp-practices/general-practice-forward-view/workforce/retained-doctor-scheme

GP Induction and Refresher Scheme Scheme for qualified GPs who are not currently on, but wish to join, the Performers' List. 2 routes:

- Refresher/returner route—for GPs who have previously been on the GMC Register and Performers List and would like to return to general practice after a >2y career break or >2y spent working abroad
- Induction route—for GPs who have qualified outside the UK and have no previous NHS experience

Separate schemes operate in England and Scotland which differ in their detail. Both schemes offer practical help, supervision/mentoring, and financial assistance including bursaries to pay for placements, and help with indemnity ± relocation costs.

Further information

Health Education England The GP Induction and Refresher Scheme. 🖱 https://gprecruitment.hee.nhs.uk/induction-refresher

NHS Scotland GP Induction and Returner Programmes. 🖱 www.scotlanddeanery.nhs.scot/your-development/gp-induction-and-returner-programmes

Career options for GPs

Times are changing in general practice. Doctors considering a life as a GP want a more flexible and varied career than in the past.

Career options within the NHS

Clinical assistant or hospital practitioner The GP works within a hospital setting on the wards or in outpatients providing a specialist service under direct supervision of a hospital consultant. Posts are usually advertised in the medical/GP press ± locally. Generally poorly paid.

GP with extended role (GPwER) Formerly GP with special interest (or GPwSI). GPs who, in addition to their normal GP duties, provide a specialist service to meet the needs of their local healthcare community: delivering a specialist clinical service beyond the scope of normal general practice, undertaking advanced procedures or developing services. The difference between a GPwER and clinical assistant is that the GPwER receives referrals from other GPs and decides on appropriate treatment independently and not under direct supervision of a consultant, but with the support of 2° care. Posts are usually advertised to local GPs. In order to be classed as having an extended role, GPs must:

- Have undertaken particular training in the specialty, or have a proven track record of expertise in the specialty. A universal accreditation process is currently being developed by the RCGP
- Regularly update knowledge through attendance at courses, conferences, or meetings and through reading
- Look after a specific group of patients with the condition
- Audit practice in the specialty area demonstrating quality of care

Medical adviser or consultant in primary care Medical advisers or directors in ambulance trusts, NHS Direct sites, etc. Some national NHS agencies have GP advisers or directors too, e.g. National Commissioning Board. Posts are either advertised or obtained by direct approach.

Providing GMS/PMS ± enhanced medical services ➲ p. 32

Providing postgraduate medical education e.g. GP tutor, GP trainer (1 in 8 GPs are GP trainers), course organizer. Approach local director or dean of postgraduate medical education.

Working for a local PCO/CCG e.g. serving on a committee, clinical tutor, GP appraiser, etc. Contact local PCO.

Opportunities outside the NHS

Academic posts A GP may be employed solely by a university or jointly by a university and the NHS. Posts include undergraduate teachers, lecturers, and research posts. Contact local university department or general practice or look for posts advertised in the medical/GP press.

Clinical sessions for commercial companies and charities e.g. school doctor for a private school.

Complementary medicine Seek specialist training. Contact representative bodies of the specialty chosen—➲ p. 128.

Forensic work e.g. police surgeon, coroner (☎ 020 8979 6805), expert witness (🖱 www.ewi.org.uk).

Media work/medical author Some sort of professional journalism qualification is useful. The BMJ offers a 1y registrar post for doctors with 3–5y experience. Courses are also available through the BMA and Medical Journalists Association (🖱 www.mja-uk.org). If you have an idea for a book, contact the medical commissioning editor of a reputable publisher to discuss your ideas.

Medical adviser posts within GP and other medical organizations e.g. RCGP, MDU, GMC. Posts may be advertised or appointments made through election or direct approach. Contact the relevant organization.

Medicals for benefits Examining Medical Practitioners (EMPs) carrying out fitness to work and disability assessments on behalf of the Department for Work and Pensions (DWP).

Medical politics GPs serve on local medical committees (LMCs) on an elected basis. Contact the local LMC and ask about standing for election.

Work for government agencies e.g. armed forces as a civilian medical practitioner or the territorial army. Usually civilian posts are advertised in the medical/GP press. For commissioned posts contact service recruitment offices.

Occupational medicine *Contact:* Faculty for Occupational Medicine 🖱 www.facoccmed.ac.uk

Prison doctor Posts are usually advertised in the medical/GP press.

Sports medicine e.g. for professional sportsmen; in private clinics. Doctors are required to have a knowledge of sports injuries, their treatment, rehabilitation, and prevention. They also need to know about other aspects of sport e.g. drugs in sport, nutrition, travel problems. *Contact*: British Association of Sport and Exercise Medicine 🖱 www.basem.co.uk

Work abroad *Contact:* RCGP International Department: E-mail: international@rcgp.org.uk; RedR UK 🖱 www.redr.org.uk; Voluntary Service Overseas 🖱 www.vso.org.uk. Overseas posts are also advertised in the medical press.

Good medical practice for GPs

GMC duties of a doctor

- Make the care of your patient your first concern
- Protect and promote the health of patients and the public
- Provide a good standard of practice and care:
    - Keep your professional knowledge and skills up to date
    - Recognize and work within the limits of your competence
    - Work with colleagues in the ways that best serve patients' interests
- Treat patients as individuals and respect their dignity:
    - Treat patients politely and considerately
    - Respect patients' right to confidentiality
- Work in partnership with patients
- Listen to patients and respond to their concerns and preferences:
    - Give patients the information they want or need in a way they can understand
    - Respect patients' right to reach decisions with you about their treatment and care
    - Support patients in caring for themselves to improve and maintain their health
- Be honest and open and act with integrity:
    - Act without delay if you have good reason to believe that you or a colleague may be putting patients at risk
    - Never discriminate unfairly against patients or colleagues
    - Never abuse your patients' trust in you or the public's trust in the profession

You are personally accountable for your professional practice and must always be prepared to justify your decisions and actions.

Reproduced with permission from General Medical Council (2006), *Good Medical Practice*, London: GMC. Available at www.gmc-uk.org/guidance

Good medical practice for GPs

- **Good clinical care** Provide best possible clinical care for patients
- **Maintaining good medical practice** Monitor, review, and continuously strive to improve performance of yourself and your practice
- **Teaching and training, appraising, and assessing** ➔ p. 8
- **Relationships with patients** Communicate with and listen to views and opinions of your patients; use terms/information they can understand; respect their privacy and dignity at all times
- **Working with colleagues** Ensure effective communication channels within/outside the practice; ensure an environment for personal/professional development for everyone working within the practice
- **Probity** Behave in a proper fashion ensuring honesty and openness in all matters. Avoid conflicts between personal and professional roles.
- **Health** ➔ p. 22

Source: data from *Good Medical Practice for GPs* (2008) www.rcgp.org.uk.

Continuity of care A patient seeing the same healthcare worker over time. In the UK this has been the norm but continuity of care is becoming less available.

Reasons for continuity of care A practitioner's sense of responsibility toward his/her patients ↑ with duration of relationship and number of contacts. Continuity builds trust, creates a context for healing, and ↑ practitioner and patient knowledge of each other. *Evidence*:

- ↑ patient and doctor satisfaction
- ↑ compliance
- ↑ uptake of preventive care
- better use of resources (time spent in the consultation, discriminatory use of laboratory tests, and admission to hospitals)

Patients' desire for personal care depends on the reason for the encounter. Most find it important to see their own GP for serious medical conditions and emotional problems.

Reasons why continuity of care is becoming less available Problems balancing accessibility, flexibility, and continuity of care:

- **Doctor factors** GP shortages, flexible careers, special interests, and managerial responsibilities limit the availability of GPs to their patients
- **Patient factors** 24h society in which patients want to be seen at their convenience rather than when their GP is available makes it impossible to maintain continuous care. For minor problems and emergencies patients do not mind who they see—as long as they see someone who can deal with their problem quickly
- **System factors** Changing roles—nurse practitioners and other healthcare professionals commonly take on tasks which used to be done by GPs; clinical governance structures mean that patients with particular conditions are managed in clinics specifically for those conditions within the practice; other primary healthcare providers, e.g. NHS 111, walk-in clinics, and separate out-of-hours cover arrangements, further fragment care

Rationing A full discussion on rationing healthcare is beyond the scope of this handbook. However, with continued innovation, rising demand, and limited resources rationing will become an increasingly important factor in medicine worldwide. To some extent there is already rationing by default—medicines and certain treatments are not provided via the NHS or have very long waiting lists. Government bodies such as NICE evaluate services and develop guidelines for healthcare professionals about medicines and services which are both clinically and cost-effective. Inevitably, this will mean that some groups will feel they are being deprived of the treatment they require. It will remain a contentious issue.

Further information

Appraisal and revalidation ➔ p. 10

GMC Duties of a doctor. 🖱 www.gmc-uk.org/guidance/good_medical_practice/duties_of_a_doctor.asp

GPC/RCGP (2008) Good medical practice for GPs. 🖱 www.rcgp.org.uk/policy/rcgp/Good_Medical_Practice_for_GPs_July_2008.ashx

The primary healthcare team

Shortages of GPs, ↑ numbers of elderly patients, and a move to increasingly specialized care being delivered in the community have changed the role of the GP. The GP has never functioned alone, but the role of the GP as co-ordinator of care in the community, directing and supporting the primary healthcare team (PHCT), has never been more important. Precise composition of PHCTs vary. Team members may include:

Practice manager General manager of the practice in liaison with the partners. *Roles include*: staff appointments, supervision, training, and dismissals; duty rotas; liaison with outside organizations (e.g. PCO) and other PHCT members (e.g. community nurses and health visitors); maintenance of premises and equipment and financial planning. Most practice managers have management qualifications.

Receptionists Perform an essential role as the interface between the general public and the GPs and nursing staff. Good interpersonal skills are essential. Training varies.

Administrative and clerical staff Perform all the non-clinical tasks necessary to keep the practice running. Training varies.

Practice nurse Duties can vary but include 'traditional' nursing tasks; health promotion; immunizations; new registration checks; specialist clinics (e.g. asthma, DM, etc.); administration and audit.

Nurse practitioner Specially trained nurse who takes on clinical responsibility for specific aspects of care he/she has been trained for either within the GP surgery or in patients' own homes, e.g. filtering out-of-hours calls or managing heart failure. Seen as a way to alleviate pressure on GPs. Nurse practitioners are at least as effective as GPs in the roles they perform.

District nurse Qualified nurse who has a community nursing qualification recognized by the Nursing and Midwifery Council. Most work is conducted in patients' homes, particularly in looking after the chronically ill or those recently discharged from hospital. District nurses are usually employed by local community trusts or PCOs and coordinate their own team of community nurses.

Community matron Highly experienced, senior nurse who works closely with a limited number of patients who are high-intensity users of health and social services (usually with serious, long-term conditions or a complex range of conditions). The community matron acts as a 'case manager' and single point of access to provide, plan, and organize care.

Health visitor Works with individuals, families, and groups in preventive medicine, health promotion, and education. Health visitors visit all babies after the midwife ceases to attend, carry out developmental assessment checks, and advise on general care and immunization. Some health visitors have a role exclusively for the elderly. Health visitors must be trained nurses and registered as health visitors with the Nursing and Midwifery Council.

Midwife Important link between hospitals, GPs, and other members of the PHCT in obstetric care. May practise independently when dealing with uncomplicated pregnancies but are obliged to refer to a doctor in the event of complications. Midwives must be registered with the Nursing and Midwifery Council.

Physician's associate (PA) Rapidly growing healthcare role in the UK. Support GPs in the diagnosis/management of patients. PAs must have undertaken a life sciences degree or other healthcare degree (e.g. nursing) before entering 2y PA training. On qualification, PAs can take medical histories, perform examinations, analyse test results, and diagnose illnesses under the direct supervision of a GP. In 2019, PAs are not able to prescribe although prescribing rights may soon be granted.

Community pharmacist Increasing role within practices—managing repeat prescribing, monitoring prescribing practices, and advising on prescribing policy.

Social worker Help people to live more successfully within the local community by helping them find solutions to their problems. Social workers tend to specialize in either adult or children's services.

Other team members Might include paramedics, dieticians, occupational therapists, physiotherapists, psychologists, and/or complementary therapists.

Social prescribing Designed to support people with a wide range of social, emotional, or practical needs. Enables healthcare professionals to refer people to a range of local, non-clinical services, e.g. volunteering, arts activities, group learning, gardening, befriending, cookery, and sports. Most models of social prescribing involve a link worker or 'navigator' who works with people to access local sources of support.

Intermediate care Community-based service working closely with the PHCT. Provided by multidisciplinary teams. Provision and team composition varies across the UK but may include specialist doctors and/or GPs, nurses, physiotherapists and occupational therapists, home carers, and social workers. Usually provided in patients' own homes but can also involve community hospitals and/or short-term nursing/residential care placements. Common service features:

- Time-limited (usually <6wk)
- Targeted at people who would otherwise face prolonged hospital stays or inappropriate admission to hospital
- Aims to maximize independence and enable people to remain living in their own homes

Further information

British Association of Social Workers 🖱 www.basw.co.uk
Faculty of Physician Associates 🖱 www.fparcp.co.uk
Nursing and Midwifery Council (NMC) ☎ 020 7637 7181 (registrations: ☎ 020 7333 9333) 🖱 www.nmc-uk.org
Royal College of Midwives 🖱 www.rcm.org.uk
Royal College of Nursing 🖱 www.rcn.org.uk

Foundation doctors in primary care

Newly qualified doctors spend their first working year (F1) doing 3 hospital placements before obtaining full registration. In the second year (F2), 42% have a 4mo attachment to primary care.

Who employs foundation doctors? Foundation programmes are hosted by hospital trusts. The trust recruits the doctors, arranges placements, employs them through their 2y programme, provides indemnity, appoints educational supervisors, and is responsible for assessment.

F2 placements in general practice

- Made by employing trusts; practices have no say about who they take
- Educational deaneries are responsible for appointing practices for F2 placements and interested practices should contact their local deanery
- Practices must be of adequate standard and have an approved supervisor for their F2s; standard required is similar to that of training practices (➔ p. 6)—practices are inspected by their deanery to check they meet the criteria. GP F2 supervisors do not need to be GP trainers. To attain approval, potential supervisors undergo a short training course; they are paid at a rate related to the trainer rate

Expectations of F2 doctors

- Do 7 clinical sessions/wk, 1 session of supervised study, 1 session of project work, and attend a half-day group session at their host trust. No OOH work is expected. Part-time working is allowed but training must be undertaken on a ≥half-time basis
- Are entitled to study leave (up to 1wk) and annual leave
- Can sign prescriptions; practical procedures must be supervised
- Need an initial induction and are likely to need longer consultation times than standard 10min slots
- Need to have a fully trained GP available whenever they are seeing patients to provide advice and support and monitor their performance

Supervised learning events (SLE) Opportunities to receive feedback from senior colleagues. 3 types are commonly used:

- Direct observation of doctor/patient encounters (minimum 3 in a 4mo placement). An experienced colleague watches the F2. ***Tools available***: Direct Observation of Procedural Skills (DOPS)—for practical procedures; Mini Clinical Evaluation Exercise (Mini-CEX)—for clinical consultations (must complete ≥2 in each 4mo period)
- Case-based discussion (CBD) Structured case review with a senior clinician (minimum 2 in any 4mo placement)
- Developing the clinical teacher—1 or more/y—designed to develop the F2's skill in teaching and/or making a presentation

E-portfolio Compulsory record of training. Includes personal reflections and SLEs. The doctor must prove for the Annual Review of Competence Progression (ARCP) that he/she has performed all the core procedures and attained all the competences required (Box 1.2) before progression to specialist training. In addition, includes Team Assessment of Behaviour (TAB), multi-source feedback, and clinical and educational supervisors' and Placement Supervision Group end-of-placement reports.

Further information

The Foundation Programme 🖱 www.foundationprogramme.nhs.uk

Box 1.2 Summary of the 4 sections of the 2016 foundation level syllabus and 20 professional capabilities

Section 1: professional behaviour and trust

1. **Acts professionally** Professional behaviour; personal organization; personal responsibility
2. **Delivers patient-centred care and maintains trust** Includes consent
3. **Behaves in accordance with ethical and legal requirements** Including confidentiality; statutory documentation; mental capacity; protection of vulnerable groups
4. **Keeps practice up to date through learning and teaching** Self-directed learning; teaching and assessment
5. **Demonstrates engagement with career planning**

Section 2: communication, team-working, and leadership

6. **Communicates clearly in a variety of settings** Communication with patients/relatives/carers; communication in challenging circumstances; complaints; patient records; interface with other healthcare professionals
7. **Works effectively as a team member** Continuity of care; interaction with colleagues
8. **Demonstrates leadership skills**

Section 3: clinical care

9. **Recognizes, assesses, and initiates management of the acutely ill patient**
10. **Recognizes, assesses, and manages patients with long-term conditions** Includes: management of long-term conditions in the acutely unwell patient; the frail patient; support for patients with long-term conditions; nutrition
11. **Obtains history, performs clinical examination, formulates differential diagnosis and management plan** Includes: history taking; physical/mental state examination; diagnosis; clinical management and review; discharge planning/summaries
12. **Requests relevant investigations and acts upon results** Investigations; interpretation of investigations
13. **Prescribes safely** Correct and clinically effective prescribing; discussion of medication with patients; guidance on prescribing; review of prescriptions
14. **Performs procedures safely** Core procedures; other procedures
15. **Is trained and manages cardiac and respiratory arrest** Including do not attempt CPR orders
16. **Demonstrates understanding of the principles of health promotion and illness prevention**
17. **Manages palliative and end-of-life care** Including care after death

Section 4: safety and quality

18. **Recognizes and works within limits of personal competence**
19. **Makes patient safety a priority in clinical practice** Patient safety; causes of impaired performance, error, or suboptimal patient care; patient identification; use of medical devices and information technology (IT); infection control
20. **Contributes to quality improvement** Quality improvement; healthcare resource management; information management

Stress in general practice

Increasing stress is a feature of society. GPs score 2× the national average on stress test scores. Similar figures are seen if anxiety scores are used and 1 in 4 GPs are classed as suffering from depression if depression screening tools are used. Burnout describes the syndrome of emotional exhaustion, depersonalization, low productivity, and feelings of low achievement. Significant numbers of GPs in all age groups are affected.

Causes of stress in general practice Insecurity about work (particularly changes in NHS structure and complaints), isolation, poor relationships with other doctors, disillusionment with the role of GPs, changing demands, work–home interface, demands of the job (particularly time pressure, problem patients, and emergencies during surgery hours), patients' expectations, and practice administration.

Roots of stress Many of the main stressors for GPs appear to be created or perpetuated by doctors' own policies: overbooking patients, starting surgeries late, accepting commitments too soon after surgeries are due to finish, making insufficient allowances for extra emergency patients, and allowing inappropriate telephone or other interruptions. Higher than average pressure scores occur in doctors with fast consultation rates compared to those with slower rates.

General characteristics of a stressed person at work Lack of concentration, poor timekeeping, poor productivity, difficulty in comprehending new procedures, lack of cooperation, irritability, aggression, withdrawal behaviour, resentment, ↑ tendency to make mistakes, and resistance to change.

Effects of stress

- **Effects on clinical work** One study showed frustrated doctors are more willing to take undesirable shortcuts in treating patients; another that those doctors with negative feelings of tension, lack of time, and frustration have poor clinical performance (measured by an ↑ prescription rate and lack of explanation to patients)
- **Effects on practices** Stress has effects on the practice too, resulting in mistakes, arguments, or angry outbursts, poor relationships with patients and staff, increased staff sickness and turnover, and accidents
- **Effects at home** Stressed GPs may develop problems in their relationships with their partners and family at home, becoming uncommunicative at home or work, and more withdrawn and isolated

Experience of stress does not necessarily result in damage. The extent of stress necessary to ↓ performance or satisfaction levels will depend on the doctor's personality, biographical factors, and coping methods but a concurrent illness or coexisting life event may have additive effects, and can ↑ vulnerability to stress or ↓ ability to cope.

Alcohol Doctors commonly use alcohol as a coping method for stress. The BMA estimates 7% of doctors are addicted to alcohol and/or other chemical substances, with half of those addicted to alcohol alone.

Interventions and solutions

- **Improve your working conditions** e.g. longer booking intervals for patient consultations; develop a specialist clinical or academic interest within or outside the practice; learn to decline extra commitments. GPs with high stress levels do not necessarily have low morale but there is a close correlation between levels of job satisfaction and morale—job satisfaction seems to protect against stress
- **Look at your own behaviour and attitudes** Stop being a perfectionist; resist the desire to control everything; don't judge your mistakes too harshly
- **Look after your own health and fitness** Set aside time for rest and relaxation; make time for regular meals and exercise
- **Allow time for yourself and your family** Do not allow work to invade family time; consider changes in working arrangements to allow more time for leisure and family
- **Don't be too proud to ask for help** As well as formal channels for seeking help, there are several informal doctor self-help organizations and counselling services (see useful contacts)

Chronic stress ➔ p. 976

Useful contacts

BMA Doctors Advisors Service and BMA Counselling Service. Provides members and their families with help, counselling, and personal support. Also produces a useful well-being webpage. ☎ 0330 123 1245 (24h) 🖱 https://www.bma.org.uk/advice/work-life-support/your-wellbeing

British Doctors and Dentists Group Support group of recovering medical and dental drug and alcohol users. Students are also welcomed. Gives confidential help and advice through a local recovering doctor or dentist. 🖱 www.bddg.org

Cameron Fund Provides a wide range of help and practical support to GPs and their dependants. ☎ 020 7388 0796 🖱 www.cameronfund.org.uk

Doctors' Support Network (DSN) Aims to raise awareness of mental health concerns, encourage doctors to look after their mental health, and to seek help early. 🖱 www.dsn.org.uk

GP Health Service For GPs/GP trainees. Confidential, self-referral NHS service for mental health concerns (including stress/depression) or addiction problems, especially where these might affect work. ☎ 0300 0303 300 🖱 http://gphealth.nhs.uk

Royal Medical Benevolent Fund Provides specialist information and advice, and necessary financial assistance due to age, ill health, disability, or bereavement. ☎ 020 8540 9194 🖱 www.rmbf.org

Sick Doctors Trust A confidential intervention and advisory service for alcohol- and drug-addicted doctors. ☎ 0370 444 5163 (24h) 🖱 www.sick-doctors-trust.co.uk

Further information

Lown M, et al. (2015) Resilience: what is it, why do we need it, and can it help us? *BJGP* **65**:e708–10.

Organizations important to general practice

British Medical Association (BMA) Voluntary professional association and independent trade union of doctors. >80% of UK doctors are members. Also runs a publishing house producing books and journals (including the BMJ); negotiates doctors' pay/terms of service; provides advice about matters related to work practice; provides educational and research facilities, accommodation, dining facilities, and financial services. The General Practitioners Committee (GPC) is a subgroup.

Further information ☎ 020 7387 4499 🖱 www.bma.org.uk

Care Quality Commission (CQC) Independent public body that:

- Assesses management, provision, and quality of health and social care in England (including GP practices—➔ p. 52)
- Regulates the independent healthcare sector through registration, annual inspection, monitoring complaints, and enforcement
- Publishes information about the state of health and social care
- Considers complaints about NHS organizations that the organizations themselves have not resolved
- Coordinates reviews and assessments of health and social care and carries out investigations of serious failures in the provision of care

Further information 🖱 www.cqc.org.uk

General Medical Council (GMC) Licenses doctors to practise medicine in the UK. It investigates complaints against doctors, and has the authority to revoke a doctor's licence if appropriate. It also monitors standards of undergraduate, postgraduate, and continuing medical education and provides information about good medical practice.

Further information 🖱 www.gmc.org.uk

General Practitioners Committee (GPC) BMA committee with authority to deal with all matters affecting NHS GPs, representing all doctors in general practice whether or not they are a member of the BMA. The committee is recognized as the sole negotiating body for general practice by the DH.

Further information 🖱 www.bma.org.uk

Local Medical Committee (LMC) Committee of GPs representative of GPs in their area. All GPs (including locums and salaried doctors) are represented by LMCs. *Functions*:

- Statutory Consultation regarding administration of primary care contracts; involvement with disciplinary and professional conduct committees; representation of GPs as a whole
- Non-statutory Advice on all matters concerning GPs; communication between GPs; links with other bodies; helping individual GPs

National Association of Sessional GPs (NASGP) Acts as a voice and resource for all NHS GPs who work independently of the traditional 'GP principal' model. This includes GP locums, retainers, salaried GPs, and GP assistants.

Further information 🖱 www.nasgp.org.uk

National Institute for Health and Care Excellence (NICE)
Special Authority aiming to provide the NHS (patients, health professionals, and the public) with authoritative guidance on ‘best practice’ and thus improve the quality/consistency of health services. It evaluates health technologies and reviews management of specific conditions.

Further information 🖱 www.nice.org.uk

Patient Advice and Liaison Service (PALS) Provided by all healthcare organizations running hospitals, GP, or community health services. Equivalent service in Scotland is the Patient Advice and Support Service (PASS). Aims to:

- Advise and support patients, their families, and carers
- Provide information on NHS services
- Listen to and record concerns, suggestions, or queries. PALS can liaise directly with NHS staff and managers regarding patients' concerns
- Help sort out problems quickly
- Direct NHS users to sources of independent advice and support

Royal College of General Practitioners (RCGP) Founded to ‘encourage, foster and maintain high standards within general practice and to act as the voice of GPs on issues concerned with education, training, research and standards’. Services include:

- Publishing—journals including *British Journal of General Practice*
- Education—online learning, courses, conferences
- Revalidation support
- Representation of GPs at national and international levels
- Support for specific groups, e.g. First5 (for GPs within 5y of their Certificate of Completion of Training); AiT Committee for GPs in training

Statutory responsibilities include:

- Developing and updating the GP training curriculum
- Setting and managing the UK licensing examination for general practice
- Managing certification and recertification

3 grades of membership:

- **Members** Are entitled to speak and vote at meetings, and to use the designation MRCGP—➔ p.7
- **Fellows** Highest grade of membership; holders use the designation FRCGP—application for Fellowship can be made by those who have been Members for >5y
- **Associates** For doctors still in training. Associates can participate in College activities but cannot vote or use the designation MRCGP

Further information 🖱 www.rcgp.org.uk

Practice in other countries

It is beyond the scope of this book to discuss different systems of healthcare and practice regulations outside the UK; however, in most countries there is a registration body (usually termed the 'Medical Council') which ensures doctors are qualified and fit to practise; an organization representing the interests of the medical profession generally (often termed the 'Medical Association'); and, separate specialist bodies representing the interests of family practitioners. Details can be obtained from the following websites:

International Directory of Medical Regulatory Authorities Lists worldwide medical regulatory bodies and contact details. 🖱 www.iamra.com

World Organization of Family Doctors (WONCA) Includes a list of member organizations and contact details. 🖱 www.globalfamilydoctor.com

European Union of General Practitioners/Family Physicians Gives overview of different healthcare systems in member states and contacts for member organizations. 🖱 www.uemo.eu

Medical Association of South East Asian Nations (MASEAN) Contains contact details for medical associations in Brunei, Cambodia, Indonesia, Laos, Malaysia, Myanmar (Burma), Philippines, Singapore, Thailand, and Vietnam. 🖱 https://masean.net/

Country-specific information

Australia

- Australian Medical Association 🖱 www.ama.com.au
- Australian Medical Council 🖱 www.amc.org.au
- Royal Australian College of General Practitioners 🖱 www.racgp.org.au

Canada

- Canadian Medical Association 🖱 www.cma.ca
- College of Family Physicians of Canada 🖱 www.cfpc.ca
- Medical Council of Canada 🖱 www.mcc.ca

China

- Chinese Medical Association 🖱 www.cma.org.cn

Hong Kong

- Hong Kong College of Family Physicians 🖱 www.hkcfp.org.hk
- Hong Kong Medical Association 🖱 www.hkma.org
- Medical Council of Hong Kong 🖱 www.mchk.org.hk

India

- Indian Medical Association 🖱 www.ima-india.org
- Medical Council of India 🖱 http://mciindia.org

Ireland (Eire)

- Irish Medical Council 🖱 www.medicalcouncil.ie
- The Irish College of General Practitioners 🖱 www.icgp.ie

Japan

- Japan Medical Association 🖱 www.med.or.jp
- Japan Primary Care Association 🖱 www.primary-care.or.jp

New Zealand

- Medical Council of New Zealand 🖱 www.mcnz.org.nz
- New Zealand Medical Association 🖱 www.nzma.org.nz
- Royal New Zealand College of General Practitioners 🖱 www.rnzcgp.org.nz

Pakistan

- Pakistan Medical and Dental Council 🖱 www.pmdc.org.pk

Singapore

- College of Family Physicians Singapore 🖱 www.cfps.org.sg
- Singapore Medical Association 🖱 www.sma.org.sg
- Singapore Medical Council 🖱 www.smc.gov.sg

South Africa

- Health Professions Council of South Africa 🖱 www.hpcsa.co.za
- South African Academy of Family Physicians 🖱 www.saafp.org
- South African Medical Association 🖱 www.samedical.org

USA

- American Academy of Family Physicians 🖱 www.aafp.org
- American Board of Family Medicine 🖱 https://www.theabfm.org
- American Medical Association (AMA) 🖱 www.ama-assn.org
- Educational Commission for Foreign Medical Graduates (ECFMG) 🖱 www.ecfmg.org
- Federation of State Medical Boards 🖱 www.fsmb.org

Chapter 2

Practice management

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GPs as managers

'If you have time to do something wrong; you have time to do it right'

W. Edwards Deming

GPs in a practice, particularly if partners, may have dual roles as both clinicians and managers of businesses.

Definition Management is the process of designing and maintaining an environment in which individuals, working together, efficiently accomplish selected aims. The manager coordinates individual effort towards the group goal. To do this, he/she needs:

- Technical skill—knowledge specific to the business of the organization
- Human skill—ability to work with people
- Conceptual skill—ability to see the 'big picture'
- Design skill—ability to solve problems

There are 5 managerial functions

- Planning Involves selecting missions and objectives and the actions to achieve them—requires decision-making
- Organizing Defining roles—ensuring all tasks necessary to accomplish goals are assigned to those people who can do them best
- Staffing Ensuring all positions in the organizational structure are filled with people able to fulfil those roles
- Leading Influencing people so that they will contribute to organization and group goals
- Controlling Measuring and correcting individual and organizational performance to ensure events conform to plans

Team work Key features which contribute to successful teamwork are:

- Communication Information sharing, feedback, and grievance airing
- Clear team rules Especially with regard to responsibility and accountability. Make sure these are understood by everyone
- Sympathetic leadership Any team needs a coordinator to direct its efforts. A weak leader may allow the team to drift; an autocratic leader may be too directive and diminish the status of other team members, and thus the effectiveness of the team
- Clear decision-making processes Especially if differences of opinion
- Pooling knowledge, experience, skills, resources, and responsibility for outcome
- Specialization of function Team members must understand and respect the role and importance of other team members
- Delegation Work of the team is split between its members. Each member leaves the others to carry out functions delegated to them
- Group support Team members share and are committed to a common, agreed purpose or goal which directs their actions

Practice meetings Essential to ensure necessary decisions are made; review policies and agree standards of care; review the financial position of the practice; educate and inform practice members; aid communication; and maintain/improve morale of practice members.

Quality improvement ➔ p. 52

Risk management Primary care is about risk and uncertainty, but sometimes unnecessary risks cause needless harm. Risk management means taking steps to minimize risk. All the major defence organizations run risk management programmes for their members. There are 4 stages:

1. Identify the risk—through analysis of complaints; comments from GPs, other practice staff, or patients; significant event review (➔ p. 54); or using defence organization data to identify common pitfalls
2. Assess frequency and severity of the risk
3. Take steps to reduce or eliminate the risk
4. Check the risk has been eliminated

Categories of risk relevant to general practice

- Clinical care, e.g. prescribing errors
- Non-clinical risks to patient safety, e.g. security and fire hazards
- Risks to the health of the workforce, e.g. hepatitis B, HIV
- Organizational risks, e.g. failure to safeguard confidential information and unlicensed use of computer software
- Financial risks, e.g. employment of a new staff member

Key safety issues for primary care

- **Diagnosis** 28% of reported errors
- **Prescribing** 1 in 5 prescriptions contains a prescribing error; 1 in 550 prescriptions contains a serious error; 9% of hospital admissions are due to potentially avoidable problems with prescribed drugs; 4% of drugs are incorrectly dispensed each year
- **Communication** Poor communication is a major cause of complaints; 28% of patients have discrepancies between the drugs prescribed at hospital discharge and those they receive in the community
- **Organizational change** Better teamwork, communication, and leadership ↓ adverse incidents

In each case, consider:

- **Organizational and management factors** Financial resources/ constraints; practice policies; organization
- **Work environment factors** Staffing levels; skill mix; work load; equipment
- **Team factors** Team structure; communication; supervision
- **Individual (staff) factors** Knowledge and skills; competence; physical and mental health
- **Task factors** Availability and use of protocols/guidelines; availability and accuracy of test results
- **Patient factors** Condition (complexity and seriousness); language and communication; personality and social factors

Change management The NHS is in constant flux and medicine doesn't stand still. Change is something that all practices have to deal with. However, change is inherently unsettling. Change management involves clear communication of the reasons for change, careful planning to ensure that everyone in the organization knows what to do, and clear leadership to maintain direction and deal with any problems that arise.

Practice contracts and payments

A number of different contracts are now available for primary care providers within the NHS. Contracting arrangements are changing rapidly and new models of care provision requiring new contractual arrangements with the NHS are likely to emerge over coming years.

General Medical Services (GMS) Contract Nationally negotiated contract between 60% of GP practices and the NHS.

Payment to practices comprises the following components
- The Global Sum +
- Enhanced Services payments +
- Payment for premises +
- IT payments +
- Quality payments +
- Dispensing payments (if applicable)

The Global Sum Major part of the money paid to practices. It is paid monthly and intended to cover practice running costs. As well as staff costs, it includes provision for delivery of:
- **Essential services** Services that all practices must undertake including: chronic disease management; general palliative care; and day-to-day medical care of the practice population (health promotion, management of minor and self-limiting illness, and referral to other services as appropriate)
- **Additional services** Services that the practice will usually undertake. Includes: certain minor surgery procedures (curettage, cautery, cryocautery of warts/verrucae and other skin lesions); child health surveillance (excluding neonatal checks); cervical screening; contraceptive services (excluding coils and implants); maternity care (excluding intrapartum care); and vaccinations and immunizations. Opting out results in a ↓ Global Sum
- **Out-of-hours care** If not opted out—➔ p. 42

Enhanced services May be agreed locally or nationally and are paid for *in addition* to the Global Sum. Examples include: more advanced minor surgery (e.g. joint injections, incisions/excisions); intrapartum care; influenza vaccination; targets for childhood immunizations; anticoagulation monitoring and minor injury services.

Payment for premises and information technology GP premises are funded in many different ways. The GP Contract has provision to reimburse practices that rent their premises the cost of the rent, or pay practices that own their premises for the use of those premises. The PCO also reimburses all the IT costs of the practice.

Quality payments Related to performance against Quality and Outcomes Framework (QOF) targets.

Dispensing Any practice in an area classified as rural may apply to dispense to patients living >1 mile from the local pharmacy, as long as this would not render the pharmacy's business unviable. A series of fees are paid for providing this service in a similar way to that in which community pharmacists are funded and in addition to the GMS Global Sum.

Other provider contracts Examples of other contractual arrangements for primary care provision currently used by the NHS include:

- **Personal Medical Services (PMS)** (40% of practices) Alternative contract for traditional GP practices. Locally agreed, and locally managed. The GMS contract has a strong influence on the content/scope of PMS Contracts but some elements of the GMS Contract may be omitted and/or additional elements may be required
- **Primary Care Led Medical Services (PCLMS)** Services delivered directly by employees of the PCO (e.g. Clinical Commissioning Group or Local Health Board)
- **Alternative Provider of Medical Services (APMS)** Services provided by any suitable individual, company or organization

Quality and Outcomes Framework (QOF) Quality incentive scheme. Data is extracted directly from practice systems. Quality indicators are reviewed and updated annually and cover both clinical and non-clinical aspects of patient care. All achievement against quality indicators converts to points, and each point achieved converts to a monetary value for the practice.

Exception reporting Prevents practices being penalized when unable to meet targets due to factors beyond their control, e.g. patients fail to attend for review or medication is contraindicated. It applies to indicators where level of achievement is determined by % of patients reaching the designated level. Practices report number of exceptions for each indicator set and individual indicator. It is important to ensure the reason why a patient has been 'excepted' is identifiable in the clinical record.

Carr-Hill Allocation Formula Geographical and social factors result in differing workload for GPs. The Carr-Hill Formula allocates Global Sum and quality payments to practices on the basis of the practice population, weighted for factors that influence relative needs and costs in order to reflect the differences in workload these factors generate. The factors included are:

- **Age and sex** Older people and children <5y require most GP care
- **Nursing and residential home residents**
- **List turnover** Areas with high list turnovers often have higher workload
- **Additional needs** Many deprived areas have higher rates of morbidity and mortality resulting in higher GP workload
- **Staff market forces** Reflects geographical variation in staff costs that practices incur across the UK
- **Rurality** Rural practices have ↑ practice costs. An additional adjustment is made for a few small practices in Scotland to allow for economies of scale (small practices incur disproportionately high costs as many expenses—particularly relating to premises—must be met regardless of practice size)

Further information

BMA The GMS Contract. 🖱 www.bma.org.uk
NHS Employers 🖱 www.nhsemployers.org

Partnership agreements

Partnership disputes are common. A properly drafted partnership agreement may prevent disputes and, if they do occur, may lessen their impact.

Partnership at will A partnership without an up-to-date written agreement is a 'partnership at will', governed by the 1890 Partnership Act. A 'partnership at will' is a very unstable situation as:

- All partners are deemed to have equal profit shares, unless there is clear evidence to the contrary
- Decisions are made by simple majority
- Notice may be served by any partner on the others without their prior knowledge or consent
- Dissolution of the partnership may take immediate effect, and no reason needs be given to justify it
- Dissolution may result in the forced sale of all partnership assets (including the surgery premises) and redundancy of all staff
- There is nothing to prevent any partner, or group of partners, from immediately forming a new practice/partnership to the exclusion of the other partner(s) once the practice is dissolved

Partnership agreements Should be drawn up every time a new partner joins or leaves a practice. Employed doctors and retainers also require contracts of employment. An agreement checklist is included in Box 2.1. Detailed guidance is produced by the BMA, and further guidance can be obtained from local BMA offices and LMCs.

Partnership disputes However good a partnership agreement is, disputes still occur. Advice on partnership and employment matters is available from the BMA and LMC, and the BMA also provides conciliation services (contact local office). Legal battles are expensive, and the BMA will not fund partnership disputes. Try to resolve matters amicably.

Discrimination It is unlawful for any partnership to discriminate on grounds of age, gender, marital status, colour, race, nationality (including citizenship), ethnic or national origins when appointing a new partner or in the way they treat an existing partner. The BMA will consider backing GPs to take such matters to industrial tribunals—contact the local office. Applications should be made on forms available via local Job Centres and must be made within 3mo of the last act of discrimination.

Contracts of employment for salaried GPs Model terms and conditions of service for a salaried GP and a model offer letter of employment are available from the BMA or DH websites. Nationally agreed salary scales apply and are compulsory for GMS, but not PMS, practices.

Practice responsibilities towards employed GPs ➔ p. 38

Box 2.1 Partnership agreement checklist

- Business detail Purpose of the business; premises and basis of occupation. If premises are owned by the partners, state procedure for valuation, payment of the retiring partner, and investment of the incoming partner
- Assets Specify assets, their ownership, arrangements for valuation and interest payments. It is illegal to sell goodwill in NHS practices
- Income and allowances Definition of practice income; allowable expenses
- Profit sharing Distribution of practice NHS income and other NHS allowances; distribution of income from non-NHS work
- Accounting Accounting and banking arrangements; bill paying; access to accounts and bank statements
- Taxation Arrangements for paying tax; obligations of each partner
- Pension arrangements
- Retirement/suspension/expulsion Reasons for suspension/expulsion; process of suspension/expulsion; mechanisms of voluntary leaving/retirement; division of assets in the event of retirement. May include a restrictive clause preventing the outgoing doctor working in the practice area for a period of time after leaving—seek legal advice
- Leave Holiday entitlement; basis of deciding who has holiday when; study leave; sabbatical leave; sick leave; maternity, paternity, and adoption leave; compassionate leave
- Obligation NHS obligations; non-NHS work within the practice; other work outside the practice; educational activities; obligations to each other; hours of work
- Decisions and disputes Decision-making process; process to manage disputes; process to dissolve partnership. Ensure that who pays legal fees for who in the event of a dispute is included
- Correct procedure Ensure each partner has signed and dated the agreement and that their signature has been witnessed. It is recommended that each partner should take independent legal advice and not rely on the ‘practice solicitor’ for sole advice

Further information

BMA 🖱 www.bma.org.uk

Equality and Human Rights Commission Equality Advisory and Support Service (EASS). ☎ 0808 800 0082; Textphone: 0808 800 0084; 🖱 www.equalityhumanrights.com

Practice premises

Funding of premises GPs may either own or rent the property in which they practise:

- **GPs who own surgeries** GPs may own surgeries by themselves or in partnership. They receive a payment ('notional rent') for allowing their private buildings to be used for NHS purposes. Payment is based on the current market rental (CMR) value of the property as assessed by the district valuer. When a new GP partner joins a practice, he/she may be expected to buy into the practice to contribute a share of previous investment in the practice premises and equipment
- **GPs who rent surgeries** Can claim reimbursement from their PCO for the rent they pay as long as it is 'reasonable' as assessed by the district valuer
- **Cost rent scheme** This scheme is no longer available, but some surgeries remain on it. Finance for building, refurbishment, or modification of GP premises was originally raised by the partners. The PCO reimburses the interest payments on the loans taken out to do this
- **Improvement grants** Available via PCOs in some circumstances

❗ New premises/refurbishments must meet national minimum standards.

Disabled access The Equality Act (2010) gives disabled people rights of access to goods, facilities, and services. A disabled person is defined as 'someone who has a physical or mental impairment that has a substantial and long-term adverse effect on his or her ability to carry out normal day-to-day activities'. Practices must:

- Not refuse to take disabled people onto a practice list or provide a lower standard of service due to their disability
- Make reasonable adjustments to their premises and the way they deliver their services so that disabled people can use them

Building regulations and access for disabled patients The building regulations exist to ensure the health and safety of people in and around all types of buildings. Part M deals with access/facilities for disabled people. All new buildings/alterations to existing buildings must be accessible to and useable by anyone, including those with disabilities.

Health and safety The basis of British health and safety law is the Health and Safety at Work Act 1974. The Act sets out the general duties employers have towards employees and members of the public, and employees have to themselves and to each other.

Responsibilities of GPs as employers The Management of Health and Safety at Work Regulations 1999 (the Management Regulations) give clear guidance about employers' duties towards their staff.

1. Employers with ≥5 employees must carry out a risk assessment and record the significant findings. HSE leaflet '5 Steps to Risk Assessment' gives more information
2. Make arrangements for implementing the health and safety measures identified as necessary by the risk assessment
3. Appoint competent people (usually the practice manager) to help implement the arrangements

4. Set up emergency procedures (e.g. fire drills)
5. Provide clear information and training to employees
6. Work together with other employers sharing the same workplace

Other important pieces of health and safety legislation

- **Employers' Liability (Compulsory Insurance) Regulations 1969** Require employers to take out insurance against accidents and ill health to their employees and display the insurance certificate
- **Health and Safety Information for Employees Regulations 1989** Require employers to display a poster, telling employees what they need to know about health and safety
- **Workplace (Health, Safety and Welfare) Regulations 1992** Cover a wide range of basic health, safety, and welfare issues, such as ventilation, heating, lighting, and seating
- **Reporting of Injuries, Diseases and Dangerous Occurrences Regulations 1995 (RIDDOR)** Require employers to notify certain occupational injuries, diseases, and dangerous events
- **Health and Safety (Display Screen Equipment) Regulations 1992** Set out requirements for work with visual display units (VDUs)
- **Personal Protective Equipment (PPE) Regulations 1992** Require employers to provide appropriate protective clothing and equipment
- **Provision and Use of Work Equipment Regulations (PUWER) 1998** Require that equipment provided, including machinery, is safe
- **Manual Handling Operations Regulations 1992** Cover moving of objects by hand or bodily force
- **Health and Safety (First Aid) Regulations 1981** Cover requirements for first aid
- **Control of Substances Hazardous to Health Regulations 2002 (COSHH)** Require employers to assess the risks from hazardous substances and take appropriate precautions
- **Gas Safety (Installation and Use) Regulations 1998** Cover safe installation, maintenance and use of gas systems and appliances in domestic and commercial premises

Computers in practices PCOs directly fund 100% of IT costs. All practices now use computers on a daily basis. All specialist GP systems must be approved by the DH (termed 'Systems of Choice'). The software covers all aspects of practice from appointment systems, through clinical care, to audit and reporting.

Electronic GP records Most GPs maintain all records on computer. *Read codes* are used to code all aspects of patient care. Characters in the code can be numerical or alphabetical. The huge number of possible combinations ensures there are enough unused Read codes to accommodate changes. *SNOMED Clinical terms (CT)* are a new coding system that will eventually replace Read coding. The aim is that all healthcare systems in the UK will use the same coding system, allowing a single unified patient record.

Further information

Health and Safety Executive 🖱 www.hse.gov.uk

Practice staff

Practices employ an array of staff. Staff costs are included in the Global Sum (➔ p. 32) paid to a practice.

Recruiting staff

- Review the post—does the post need to be filled or the duties changed?
- Prepare a job description stipulating duties and hours of work
- Prepare a profile of the person required
- Decide on a salary range; the BMA can give advice
- Advertise the post
- Set a closing date for applications
- Shortlist candidates
- Interview—decide who will interview, what points must be covered, and who will ask questions; ask similar questions to all candidates, and score the responses at the time
- Make a decision on the preferred candidate—if in doubt, defer the appointment or re-interview preferred candidates
- Confirm the job offer by letter asking for a formal written acceptance by email or letter in return
- Plan an induction course for the new employee; a probationary period can be helpful for both employer and employee
- Produce a contract of employment

Employment law Very complex field which changes rapidly. If in doubt, contact your local BMA office for advice. Major points:

Contract of employment Sample contracts are available from the BMA. Employees have a contract of employment from the day they accept their job—even if it is not written. All employees must be provided with a written statement of the main particulars of their employment <2mo after their start date. This must include: pay, hours, holidays, notice period, disciplinary and grievance procedures.

Pay Workers must be paid at least the national minimum wage for every hour worked. Deductions can only be made if authorized by legislation, contract of employment, or in advance in writing by the employee. All employees must receive an itemized pay statement at, or before, the time they are paid, including all deductions.

Notice After 1mo employment, an employee must give ≥1wk notice. An employer must give an employee ≥1wk notice after 1mo, 2wk after 2y, 3wk after 3y, and so on up to 12wk after ≥12y, unless longer notice periods are specified in the contract of employment.

Redundancy pay After 2y continuous employment, employers must make 'redundancy payments' related to employee's age, length of continuous service with the employer (to a maximum of 20y), and weekly pay.

Time off Employees are entitled to time off for illness; antenatal care; emergencies involving a dependant; certain public duties (e.g. jury service); to look for another job; and for approved trade union activities.

Working time Parents of children <6y or disabled children <18y and carers may request flexible working patterns; employers have a duty to consider their requests. Working Time Regulations (1998) apply to agency workers and freelancers as well as employees and include:

- Average working week ≤48h (although individuals can opt to work longer)
- A minimum of 1d off each week
- A minimum of 5.6wk paid annual leave (28d for a full-time worker)
- 20min in-work rest break if the working day is ≥6h
- 11 consecutive hours' rest in any 24h period (night workers must work ≤8h/d)

Pensions All employees must belong to a pension scheme. The NHS pension scheme is available to practice employees.

Maternity leave All pregnant employees are entitled to 52wk maternity leave (26wk ordinary maternity leave + 26wk additional maternity leave) regardless of length of service. Women are entitled to return to their own or an equivalent job after their leave. Similar arrangements are in place for adoptive mothers.

Paternity leave Employees who have worked for their employer for ≥26wk by the 15th wk before the baby is due and up to the birth of the child are entitled to 1–2wk paternity leave which must be completed within 56d of the birth. Fathers may also claim additional paternity leave for up to 26wk from 20wk to 1y after birth/adoption to look after their child if the mother returns to work.

Parental leave After 1y employment, employees are entitled to 13wk unpaid parental leave for each child born or adopted up to the child's 5th birthday (or 5y after adopted). Parents of disabled children can take 18wk up to the child's 18th birthday.

Health and safety of staff ➔ p. 36

Discrimination Employers must not, either directly or indirectly, discriminate against their staff on the basis of age, race, gender, or disability.

Unfair dismissal Employees of >1y standing (or on maternity, paternity, or adoption leave) are entitled to a written statement of reasons for dismissal. Employers must not dismiss an employee unfairly.

Further information

ACAS Provides advice for employers and employees, downloadable employment tools and checklists, free e-learning on employment matters and an advice helpline. ☎ 0300 123 1100 🖱 www.acas.org.uk

Equality and Human Rights Commission Equality Advisory and Support Service (EASS). ☎ 0808 800 0082; Textphone: 0808 800 0084; 🖱 www.equalityhumanrights.com

HM Government Information about 'Employing people'. 🖱 www.gov.uk

Registration of patients

Practice area and boundaries ➔ p. 4

Practice leaflets Each practice is required to produce a practice leaflet to distribute to patients. In Wales, the practice leaflet must be in Welsh and English. The practice leaflet informs patients about the practice, the services provided, and how to access them. In addition it informs patients of their rights and responsibilities. Most practices also include general health information and information about self-management of minor illness.

Registration process Patients can apply to join a practice list by handing in their medical card at the practice or completing an application form. For children, a parent or a guardian can make the application.

- **Open lists** Practices with open lists must consider all applications to join their list
- **Closed lists** Practices with closed lists can only consider applications from immediate family members of patients already registered

List closure Practices wishing to close their lists must inform the PCO in writing. The PCO must then enter into discussion with the practice to provide support to keep the list open. If that is not possible, the list will be closed for a specified period of time or until the list size falls below a set limit.

Newly registered patients When a patient has been accepted onto a practice list, the practice must offer the patient a consultation for a routine health check (the 'new patient check') within 6mo of registration.

Temporary residents Patients may register with a practice on a short-term basis for treatment or advice if they are living temporarily (for >24h but <3mo) in the practice area.

Emergency and immediately necessary treatment Practices must provide services required in core hours for the treatment of anyone:

- Injured or acutely unwell at any place in its practice area
- Whose application for inclusion in the practice list (as a permanent or temporary resident) has been refused and who is not registered with another provider in the area

Assignments PCOs may assign patients to any open practice list if the patient has problems registering with a practice. PCOs can only assign patients to closed lists if all other local practice lists are also closed.

Removing patients from the practice list Practice policies for removing patients from the practice list should be stated in practice leaflets.

Situations that justify removal

- **Violence** Physical violence or verbal abuse towards doctors, practice staff, premises, or other patients by patients *or* other household members
- **Crime and deception** e.g. deliberate deceit to obtain a service or benefit; obtaining drugs under false pretences for non-medical reasons; stealing from practice premises
- **Relocation to an address out of the practice area** Unless the practice is participating in the Patient Choice Scheme ➔ p. 4

Situations that never justify removal

- Costly or difficult treatment
- Age General practice is about looking after patients from cradle to grave. Although patients >75y do result in higher costs, this is reflected in allocation of funds to the practice

Situations that do not normally justify removal

- Disagreement with the patient's views Patients must have freedom to choose whether to accept a GP's advice
- Critical questioning and/or complaints Complaints via normal in-house channels can be constructive and help improve services; they do not usually justify removal from the practice list. However, personal attacks on a doctor or allegations that are clearly unfounded indicate a serious breakdown in the doctor–patient relationship and could justify removal

Other family members Removal of other family members should not automatically follow removal of a patient from a practice unless ongoing care of the rest of the household would be impossible.

Removing patients from the practice list

- Warn the patient Patients must have received a warning and explanation for the reasons why a practice is considering an application for removal from the practice list <12mo before an application for removal is made to the PCO. Exceptions to this rule are violent patients, patients who have moved outside the practice area, and those for whom it would be unsafe or impractical to issue a warning
- Inform the PCO in writing of your decision Except in the case of violent patients, removal will not take effect until the 8th day after the request is received by the PCO unless the patient is accepted by another GP. The patient will be notified by the PCO
- Write to the patient about the decision and reason for removal (take advice from your medical defence organization, if needed). Include information on how to register with another practice and reassurance that the patient will not be left without a GP. Take care to ensure reasons given are factual and the tone of the letter is polite/informative

Immediate removal of violent patients

- Notify the police (or, in Scotland, either the police or the procurator fiscal) about the violent behaviour
- Notify both the PCO and the patient of the removal in writing The PCO has a duty to provide alternative primary medical care services by commissioning specialized directed enhanced services, e.g. GPs with secure facilities for consulting

Patients' rights to change doctor Patients also have a right to change their doctor. They are not required to give reasons or any period of notice, and there is no requirement for the GP to be notified.

Eligibility for free healthcare Eligibility for NHS care is likely to change once the UK leaves the European Union. However, all emergency care is likely to remain free of charge to all patients regardless of country of origin or residence entitlement, and whether delivered in 1° or 2° care.

Out-of-hours services

What is urgent care? Urgent care refers to the range of responses that the health and care services provide to people who need, or perceive that they need, urgent advice, care treatment, or diagnosis. Urgent 'same day' appointments currently account for 1 in 3 daytime consultations in general practice, so all GPs must have the skills/knowledge to manage patients with a perceived urgent care need. Effective GPs can manage patients with urgent care needs so that 'dangerous diagnoses' are not missed and A&E and acute hospital services are used efficiently.

What are OOH services? Urgent primary medical care is currently provided by out-of-hours (OOH) services from 6.30 p.m. to 8 a.m. on weekdays and throughout weekends and public holidays—a total of at least 70% of every week. In England alone, every year primary care OOH services:

- Receive 8.6 million calls
- Complete 6.8 million assessments—3 million in OOH primary care centres (44%), 2.9 million by telephone (43%), and 0.9 million through home visits (13%)
- 1.5% of the calls dealt with are considered 'life-threatening' emergencies and 15% are classified as 'urgent'

'Opting out' of OOH Both PMS and GMS practices can 'opt out' of providing an OOH service. The decision must be made for the whole practice; individual doctors within a practice cannot 'opt out' alone. The cost of opting out is 7% of the Global Sum (or PMS equivalent).

OOH work by 'opted out' practices There is nothing to stop practices that have opted out from offering surgeries or consultations within the time periods specified as OOH. These services can be paid for through the practice Global Sum or under the 'extended opening' arrangements with local commissioners. With government promises of extended day-time and weekend availability of routine GP services, and emergence of new models of care, the division between within-hours urgent care provision and OOH care is becoming increasingly blurred.

OOH and urgent care providers PCOs can consider a range of providers as long as accreditation standards are met. Only where a practice is exceptionally remote, is the PCO able to require a practice to provide OOH care. Special arrangements for payment then exist. Several schemes currently operate side by side:

- **In-practice rota** Traditional model of cover that is now very rare; usually organized in a rota between practice GPs; largely based on home visiting
- **Extended rota** GPs on-call in rotation for a small group of practices. Some extended rotas still operate in rural areas
- **GP cooperative** GPs within an area grouped together (often >100 practices in a co-op or federation) to cover urgent/OOH care either between themselves or by employing other GPs and allied health professionals; often several individuals are 'on call' at any time, e.g. one doing visits; one taking calls; one seeing patients in a clinic, etc.

- PCO or commercial urgent and/or OOH care provider Employ GPs, paramedics, physician's associates, and specialist nurses to provide both surgery-based and home visiting services
- Hospital-based OOH/urgent care clinics GPs, advanced nurse practitioners/physician associates, and primary care nurses attached to A&E
- NHS walk-in centres Walk-in clinics tend to offer nurse consultation and use NHS diagnosis and management algorithms; most are sited in urban areas and aim to provide easier access to medical care

❗ Since 2014, GP practices in England have a responsibility for monitoring the quality of OOH services for their registered patients.

NHS 111 and NHS 24 24h nurse-led telephone advice service available throughout England (NHS 111), Wales (NHS 111 Wales), and Scotland (NHS 24). Provides a single point of contact for patients requiring urgent medical advice or treatment; telephone services are supported by websites providing healthcare information.

Challenges for healthcare practitioners

- There is a higher proportion of very ill patients in urgent and OOH settings; healthcare practitioners may need to commence resuscitation or critical illness management protocols more frequently
- Despite better sharing of medical records, there is often no information about patients apart from the information that the patient/carer provides when a patient is seen in an urgent or OOH setting
- Team size: OOH and urgent care providers often have a much larger team than healthcare practitioners may be used to when working in traditional general practice. Workers may not know other team members and may be unfamiliar with their working environment
- Healthcare practitioners may work alone for a high proportion of their time doing home visits or manning an OOH clinic
- Access to drugs and some services may be limited
- First contact is almost always over the telephone and good communication skills are needed to provide accurate assessment, triage to appropriate care, and ensure safety netting

Pre-hospital emergency care To meet calls for increasing sophistication of pre-hospital emergency care, a new subspecialty in that field has been created. For GPs, particularly those working in urgent care settings or with populations that are geographically remote from acute specialist services, expertise in this subspecialty would clearly be beneficial. However, currently GP training does not allow GPs to meet the basic entry requirement for this subspecialty. Changes in GP training in the future may remedy this.

Further information

NHS 111 ☎ 111 🖱 www.nhs.uk
NHS 24 ☎ 111 🖱 www.nhs24.scot
NHS Direct Wales ☎ 0845 4647 🖱 www.nhsdirect.wales.nhs.uk

Patient records

General principles Patient records should be factual, consistent, and accurate. Ensure a logical sequence; be clear, unambiguous, and concise. Use standard coding techniques if using an electronic record—standard templates may help. Wherever possible, write notes openly while patients/carers are present in terms they can understand. Ensure that the record is correctly dated, timed, and identifiable to the person creating the record. *Record*:

- **Information on which you have based your decisions** Presenting problems; past/family history; examination findings and test results
- **Your impression of the situation** How you see the problem—may include diagnosis, differential diagnosis, prognosis
- **Plan of action** Negotiated between patient and doctor—may include tests requested, prescriptions given, referrals made
- **Information shared and advice given** Relevant worries/concerns voiced by the patient; information/advice provided to the patient—especially safety netting advice and review/follow-up arrangements
- **Other essential information** e.g. correspondence to/from other agencies; if consent for treatment/examination (➔ p. 48) was given

❗ *Do not include* Abbreviations (especially unconventional ones); jargon; or personal views about behaviour or temperament unless they have a bearing on the management of the patient.

Confidentiality ➔ p. 46 **Electronic patient records** ➔ p. 37

Summary Care Record (SCR) Electronic medical notes summary extracted from GP IT systems that can be viewed by healthcare staff in other NHS settings. At a minimum, the SCR holds information about current medication, and allergies/adverse reactions to medicines. The patient can also choose to include additional information, e.g. long-term conditions, significant medical history, or specific communication needs.

Shared electronic GP records NHS plans for healthcare organizations to work more closely together often depend on sharing patient information, including GP records. As a result, local data sharing arrangements are in place throughout the UK. However, the registered GP practice legally remains the ‘data controller’ for the record.

- Be clear on the sharing arrangements that you are part of
- Communicate and explain data sharing arrangements to patients; gain explicit consent for data sharing where possible and record any objections. Some GP systems allow patients to view who has looked at their data
- Act on any indications of illegitimate record access

Patient Online Allows patients online access to practice systems and their electronic GP notes to book/cancel appointments, order repeat prescriptions, and view their GP record including test results and hospital correspondence.

Amending records Rectify errors of fact or judgement. Any alterations or additions should be dated, timed, and signed in such a way that the original entry can still be seen. Patients may request correction of information they believe is incorrect—you must record the patient’s view. Highlight amendments and reasons for them.

Subject access requests (SARs) Under the Data Protection Act 1998 and 2018, patients have a right of access to health records which:

- Are about them and from which they can be identified
- Consist of information relating to their health or condition
- Have been made in connection with their care

Requests should usually be written (paper or electronic). Medium in which records are supplied can be agreed between the patient and the practice and may be paper (photocopy, computer printout), electronic (e.g. USB stick), or by facilitating patient access to Patient Online.

Since May 2018, practices cannot charge a fee for this service, although may charge for repeat requests. The entire record should be provided within 1mo of the SAR unless the patient agrees to limit the request (e.g. to electronic records only, or records made since a defined date).

Who can seek access?

- Any competent person may seek access to their own health records, including competent children (➔ p. 46)
- Any person with parental responsibility may apply for access to records of a child (<18y, or <16y in Scotland). Where >1 person has parental responsibility, each may apply independently without consent of the other parent
- A third party authorized by a competent person may seek access to that person's records (e.g. solicitor or insurance company), but proof of permission from the patient must be provided. If there is doubt, contact the patient to verify consent has been given

Mentally incapacitated adults Where access is sought and the individual lacks capacity to give permission, decisions must be based on the person's best interests, taking into account the views of the person's representative(s) and the individual's expressed wishes and values—➔ p. 46.

Access to dead patients' records ➔ p. 46

❗ If unsure, take advice from the BMA or your defence organization.

Security of records Do not leave records (electronic or manual) unattended in easily accessible areas. When not in use, store paper files and portable electronic equipment locked up. Query the status of strangers. Highlight concerns to the practice manager. Do not reveal how security systems work.

- **Manual records** Store files closed and in logical order. Use a tracking system to monitor whereabouts of files, and return files as quickly as possible
- **Electronic records** Do not leave terminals unattended and logged in. Do not share logins or reveal passwords to others. Change passwords regularly, and avoid using short or obvious ones. Keep smart-cards securely. Always clear the screen of a previous patient's information before seeing another. Use a password-protected screen saver to prevent casual viewing of patient information by others

Further information

BMA 🖱 www.bma.org.uk

- Access to health records: guidance for health professionals in the UK
- Medical records access: GDPR changes to Subject Access Requests and fees from 25 May 2018

GMC Guidance on good practice—confidentiality. 🖱 www.gmc-uk.org

Confidentiality

Respect for confidentiality is also an essential requirement for the preservation of trust between patient and doctor. Failure to comply with standards can lead to disciplinary proceedings and even restriction/cessation of practice.

Caldicott Principles for disclosure of patient information

- **Justify the purpose** Patients may agree to identifiable information about themselves being released to specific individuals for known purposes. Implied consent applies when patients are aware that personal information may be shared and of their right to refuse but make no objection. Patients must have a realistic opportunity to refuse—and if they do refuse, clearly document that and respect their decision
- **Do not use patient identifiable information unless it is absolutely necessary** It is not necessary to seek consent to use anonymous information. If in doubt, seek advice from the BMA or your defence organization. Health information used for secondary purposes, e.g. planning, teaching, audit, should—when possible—be anonymous
- **Use the minimum patient identifiable information**
- **Access to patient-identifiable information should be on a strict 'need-to-know basis'**
- **Everyone should be aware of their responsibilities**
- **Understand and comply with the law**

Special circumstances

Children (<16y) Disclosure can be authorized by a person with parental responsibility. Young people, mature enough to understand the implications, can make their own decisions and have a right to refuse parental access to their health record.

Mentally incapacitated adults Assessment of capacity to consent to information disclosure is time- and decision-specific. A mentally incapacitated adult can consent to information disclosure if the person is able to:

- Understand the concept of authorizing/prohibiting sharing of information
- Retain that information long enough to make a decision
- Weigh up the implications of disclosure or non-disclosure
- Communicate a decision

Otherwise, decisions must be based on an evaluation of the person's best interests, taking into account the views of the patient's representative(s) and reflecting the individual's expressed wishes and values.

! Except in Scotland, parents are able to consent for mentally incapacitated 16–17y olds.

The deceased Legislation covering records made since 1 November 1991 permits limited disclosure in order to satisfy a claim arising from death. Where there is no claim, there is no legal right of access to information.

Breaching confidentiality Only breach confidentiality in exceptional cases and with appropriate justification. This includes discussing a patient with another health professional not involved currently with that patient's care. Wider disclosure to people loosely associated with care (e.g. support staff in residential care settings) requires patient consent.

Situations where breach of confidentiality may be justified

- **Emergencies** Where necessary, to prevent or lessen a serious and imminent threat to the life or health of the individual concerned or another person (unless previously forbidden by the patient)
- **Statutory requirement** Ask under which legislation it is sought—check the legislation before disclosing if unsure
- **The public interest** What is in the public interest is not defined. The BMA has produced guidance
- **Public health** Reporting notifiable diseases (statutory duty)
- **Required by court or tribunal**
- **Adverse drug reactions** Routine reporting to the Medicines and Healthcare products Regulatory Agency (➲ p. 120)
- **Complaints** As part of GMC performance procedures involving doctors

Legal considerations

- **Human Rights Act (1998)** Establishes a right to 'respect for private and family life' and creates a general requirement to protect the privacy of individuals and preserve confidentiality of their health records. Compliance with the Data Protection Act and common law of confidentiality should satisfy requirements
- **Common law of confidentiality** Built up from case law where practice has been established by individual judgements. The key principle is that information confided should not be used or disclosed further, except as originally understood by the confider, or with their subsequent permission, except in exceptional circumstances (➲ p. 46)
- **Administrative law** The extent the NHS can access confidential information to perform its functions is set down in statutes
- **Health and Social Care Act (2001)** Allows for certain exceptions to confidentiality laws to be made, e.g. for use in cancer registries
- **Freedom of Information Act (2000)** Applies to all NHS bodies, including GP practices. Practices are required to produce a publication scheme detailing all information routinely published by the practice. In addition, members of the public can make written requests to see any information recorded by the practice in any format. These rights are restricted by certain exemptions, e.g. personal data
- **Data Protection Act (1998 and 2018)** Imposes constraints on processing of personal information. Also requires personal data to be protected against unauthorized/unlawful processing and accidental loss, destruction, or damage. Also applies to personnel records
- **European General Data Protection Regulation (GDPR) (2018)** European legislation strengthening individual rights to control of data, to be informed of any data breaches and creating a right to 'erasure'

Further information

BMA Confidentiality and people under 16. 🖱 www.bma.org.uk
GMC Guidance on good practice—confidentiality. 🖱 www.gmc-uk.org
Information Commissioner's Office Data protection. 🖱 www.ico.gov.uk

Consent

Consent Implies willingness of a patient to undergo examination, investigation, or treatment (collectively termed 'procedure' in this section). It may be expressed (i.e. specifically says yes or no/signs a consent form) or implied (i.e. complies with the procedure without ever specifically agreeing to it—use with care). For consent to be valid, patients:

- Must be competent to make the decision
- Have received sufficient information to take it
- Not be acting under duress

Under 'common law', touching a patient without valid consent may constitute the civil or criminal offence of battery, and if the patient suffers harm as a result of treatment, lack of consent may be a factor in any negligence claim. Never exceed the scope of the authority given by a patient, except in an emergency.

If you are the doctor carrying out a procedure, it is your responsibility to discuss it with the patient and seek consent. The task may be delegated, but the responsibility remains yours.

Information to include

- Reasons why you want to perform the procedure
- Nature, purpose, and side effects (common and serious) of proposed procedure
- Name of the doctor with overall responsibility
- Whether students or other 'trainees' will be involved
- Whether part of a research programme or outside usual procedure
- Reminder that patients have a right to seek a second opinion and/or can change their minds about a decision at any time

And for therapeutic procedures/treatments

- Details of diagnosis and prognosis (including uncertainties)
- Management options—including the option not to treat and other options that you cannot offer—and, for each option, an estimation of likely risks, benefits, and probability of success
- Details of follow-up in order to monitor progress or side effects

❗ Document if a patient doesn't want to be fully informed before consenting.

Written consent It is good practice to seek written consent if:

- The procedure is complex or involves significant risks ('risk' means any adverse outcome, including complications and side effects)
- The procedure involves general/regional anaesthesia or sedation
- Providing clinical care is not the primary purpose of the procedure
- It has consequences for employment, social, or personal life of the patient
- The procedure is part of a project or programme of approved research

Establishing capacity to make decisions ➔ p. 96

Mentally incapacitated adults The Mental Capacity Act (2005), and equivalents in Scotland and Northern Ireland, enable patients' advocates (usually friends, relatives, or carers) or suitable professionals (e.g. doctors, social workers) to act in patients' best interests on their behalf. This includes provision of medical care. Before acting:

- Take all factors affecting the decision into consideration
- Involve the patient with the decision-making as far as possible
- Take the patient's previous known wishes into consideration
- Consult everyone else involved with the patient's care/welfare

In situations in which there is disagreement about the patient's best interests, the decision can be referred to the Court of Protection.

Advance statements ➔ p. 97

Children (<16y) A competent child is able to understand the nature, purpose, and possible consequences of a proposed procedure as well as the consequences of not undergoing that procedure. This is termed 'Gillick competence' after the court case in which the principle was established (Gillick *v* West Norfolk and Wisbech AHA [1986] AC 122).

A competent child may consent to treatment. However, if treatment is refused, a parent or court may authorize procedures in the child's best interests.[1] Where a child is not judged competent, only a person with parental responsibility may authorize/refuse investigations or treatment. If in doubt, seek legal advice.

Emergencies When consent cannot be obtained, you may provide medical treatment, provided it is limited to what is immediately necessary to save life or avoid significant deterioration in the patient's health. Respect the terms of any advance statement/living will you are aware of.

Further information

GMC (2008, under review) Consent: patients and doctors making decisions together. 🖱 www.gmc-uk.org/ethical-guidance/ethical-guidance-for-doctors/consent

GMC (2018) 0–18 years: guidance for all doctors. 🖱 www.gmc-uk.org/ethical-guidance/ethical-guidance-for-doctors/0-18-years

Office of the Public Guardian Health and social care workers: Mental Capacity Act decisions. 🖱 www.gov.uk/government/publications/health-and-social-care-workers-mental-capacity-act-decisions

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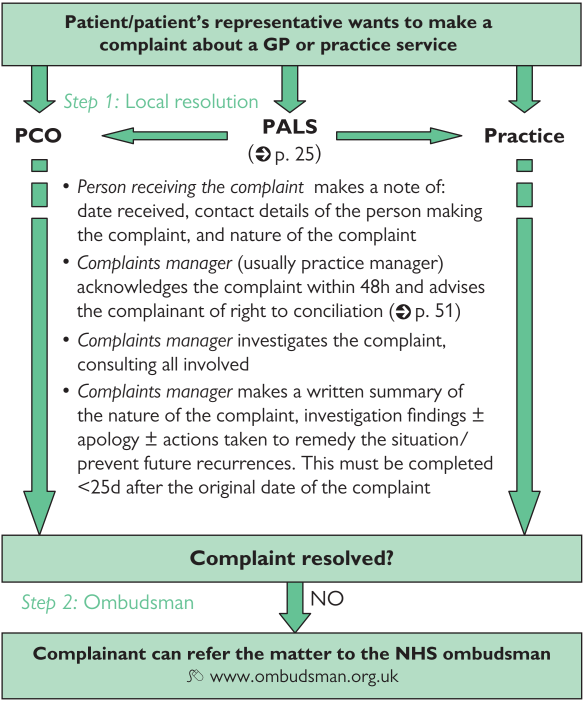
[1] *Note*: in Scotland, parents do not have this power to overrule a competent child's decision.

Complaints

Sadly, complaints are a fact of life for most GPs. The most constructive and least stressful approach is to view them as a learning experience and a chance to improve practice risk management strategy. Always contact your local LMC ± defence organization if you are directly implicated in a complaint. Patients who complain generally want:

- Their complaint to be heard and investigated promptly
- Their complaint to be handled efficiently and sympathetically
- To receive a genuine apology if mistakes have occurred
- To be assured that steps will be taken to prevent a recurrence

NHS complaints procedure for general practice Figure 2.1


Patient/patient's representative wants to make a complaint about a GP or practice service
Step 1: Local resolution
PCO
PALS
(➲ p. 25)
Practice
Complaint resolved?
Step 2: Ombudsman
NO
Complainant can refer the matter to the NHS ombudsman
🖱 www.ombudsman.org.uk

- *Person receiving the complaint* makes a note of: date received, contact details of the person making the complaint, and nature of the complaint
- *Complaints manager* (usually practice manager) acknowledges the complaint within 48h and advises the complainant of right to conciliation (➲ p. 51)
- *Complaints manager* investigates the complaint, consulting all involved
- *Complaints manager* makes a written summary of the nature of the complaint, investigation findings ± apology ± actions taken to remedy the situation/prevent future recurrences. This must be completed <25d after the original date of the complaint

Figure 2.1 The NHS complaints procedure for general practice

Time limits for complaints NHS complaints can only be accepted:

- <1y after the incident which is the subject of the complaint, or
- <1y after the date at which the complainant became aware of the matter

After that time, complaints can only be accepted if there is good reason for delay and it is possible to effectively investigate.

A 3y time limit after the incident (or after the date upon which the claimant became aware that the incident might have caused harm) is placed on civil clinical negligence cases, except for children who may claim until their 21st birthday.

Conciliation Is a way of dealing with complaints that helps to avoid adversarial situations. Either party can ask the local PCO for conciliation, but both parties must agree to it taking place. By bringing the two sides together with a neutral conciliator, it aims to:

- Explain and clarify matters for both parties
- Ensure both parties are really listening to each other
- Ensure the process is unthreatening and helpful

Records of complaints A file on the complaint, including a copy of all correspondence, should be kept separate from clinical records of the patient and, if the patient leaves the practice, should not be sent on with the clinical notes.

Private sector Most private sector healthcare providers have their own complaints resolution procedures. Patients should contact the organization concerned for details.

Disciplinary procedures There is no direct connection between complaints procedures and disciplinary action. If a complaints procedure reveals information indicating the need for disciplinary action, it is the responsibility of the PCO to act. If they decide there has been a breach of the terms of service, the PCO can fix a penalty, if appropriate.

Further information

Risk management ➔ p. 31
BMA 🖱 www.bma.org.uk
Medical defence organizations

Quality improvement

Quality improvement (QI) comprises a set of values and tools for proactively setting goals and planning, implementing, and measuring change in order to improve patient care. A culture of quality should exist throughout all healthcare organizations, and every team member should be involved in delivering and improving quality.

Steps to quality improvement Establish a quality improvement culture. Within practices there should be continuous review and appraisal of procedures and standards—RAID:

- Review—gather all stakeholders together to look at a topic
- Agree—a strategy to take forward. All objectives should be SMARTS (specific, measurable, achievable, relevant, time limited, and sustainable)
- Intervene—make changes decided upon
- Demonstrate—the effect of changes through audit (➔ p. 54), patient satisfaction questionnaires, prescribing data, etc.

Methods of identifying topics for QI Tools include:

- Review of new guidelines/clinical pathways for compliance
- 'Always events'—feasible care processes that patients identify should always happen when they interact with GP practices, e.g. timely appointments
- 'Never events' (Box 2.2)—a validated list of events known to cause severe harm that are completely preventable—could any of these events happen in your practice? How might you minimize risk?
- General Practice Safety Checklist (MoRRIS checklist)
- Trigger tool—review a sample of records of high-risk patients (e.g. patients who are designated 'frail') for undetected incidents
- External peer review of practice procedures and care pathways
- Significant event reviews—➔ p. 54
- Criterion-based audit—➔ p. 54

Risk management ➔ p. 31

Clinical governance Defined as 'a framework through which organizations are accountable for continuously improving the quality of their services and safeguarding high standards of care by creating an environment in which excellence in clinical care will flourish'.

Essential elements of clinical governance Figure 2.2

Care Quality Commission (CQC) The Health and Social Care Act 2008 required any individual, partnership, or organization providing healthcare services in the UK to register with CQC as a service provider.

Monitoring After registration, CQC regularly inspects practices to ensure that they meet essential standards. After inspection, each practice is rated according to its performance against CQC criteria and awarded an overall rating of 'Outstanding', 'Good', or 'Inadequate'.

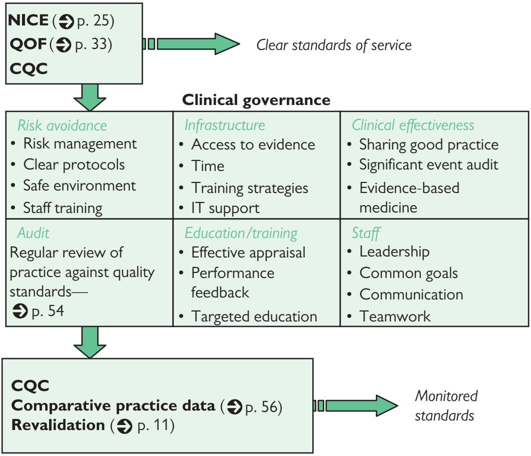
❗ Practices deemed inadequate by CQC usually remain registered but must ensure that they take steps to manage any risks posed by non-compliance, and must submit an action plan to explain the steps that will be taken to meet compliance with a target date for achievement.

Concerns about GP performance ➔ p. 11

Box 2.2 'Never events' for QI

- Drug prescribed to a patient where it is recorded in the patient's notes as having previously caused a severe adverse reaction
- Planned referral of a patient with suspected cancer is not sent
- Prescribing a teratogenic drug to a patient known to be pregnant (unless initiated by a specialist)
- Emergency transport is not discussed or arranged when admitting a patient as an emergency
- Abnormal investigation result is received but not clinician reviewed
- Aspirin is prescribed for a <12y-old patient (unless recommended by a specialist for a specific clinical condition, e.g. Kawasaki's disease)
- Systemic oestrogen-only hormone replacement therapy is prescribed for a patient with an intact uterus
- Methotrexate is prescribed daily rather than weekly (unless initiated by a specialist for a specific clinical condition, e.g. leukaemia)
- Needle-stick injury is caused by a failure to dispose of 'sharps' in compliance with national guidance and regulations
- Adrenaline (or equivalent) is *not* available when clinically indicated for a medical emergency in the practice or GP home visit

Reproduced from de Wet et al. Developing a preliminary 'never event' list for general practice using consensus-building methods. *BJGP*, **64** 620), e159–e167, 2014. Copyright © British Journal of General Practice 2014. Distributed under the terms of the Creative Commons Attribution 3.0 Unported (CC BY 3.0) http://creativecommons.org/licenses/by/3.0/


NICE (➲ p. 25)
QOF (➲ p. 33)
CQC
Clear standards of service
Clinical governance
CQC
Comparative practice data (➲ p. 56)
Revalidation (➲ p. 11)
Monitored standards

Risk avoidance	*Infrastructure*	*Clinical effectiveness*
• Risk management • Clear protocols • Safe environment • Staff training	• Access to evidence • Time • Training strategies • IT support	• Sharing good practice • Significant event audit • Evidence-based medicine
Audit	*Education/training*	*Staff*
Regular review of practice against quality standards—➲ p. 54	• Effective appraisal • Performance feedback • Targeted education	• Leadership • Common goals • Communication • Teamwork

Figure 2.2 How does clinical governance fit with other QI initiatives?

Further information

Bowie P, et al. (2015) Participatory design of a preliminary safety checklist for general practice. *BJGP* **65**(634):e330–43.
Care Quality Commission 🖱 www.cqc.org.uk
de Wet C, et al. (2014) Developing a preliminary 'never event' list for general practice using consensus-building methods. *BJGP* **64**(620):e159–67.

Audit and research

Audit is defined as the systematic critical analysis of quality of healthcare. Its purpose is to appraise current practice (*What is happening?*) by measuring it against pre-selected standards (*What should be happening?*), to identify and implement areas for change (*What changes are needed?*) and thus improve performance. Audit is a continual process and an integral part of quality improvement activity (➔ p. 52).

Criterion-based audit: the audit cycle The process of identifying areas of care to be audited, implementing necessary changes, and periodically reviewing the same issues is known as the audit cycle.

- **Choosing a topic** Any practice matter—clinical or administrative. Make sure the topic is important, manageable, clearly defined, and data is available to assess the criteria chosen. Good starting points: significant events, QOF targets, complaints, clinical guideline topics, and personal observations
- **Choosing criteria** Criteria are specific statements of what should be happening. Criteria might be those laid down for quality payments, 'gold standard' care as defined in guidelines, or generated within the practice. Use evidence-based criteria wherever possible. All criteria have to be measurable—ideally with data already collected
- **Setting standards** Standards are minimum levels of acceptable performance for a criterion. 100% achievement of standards is unusual so set realistic standards based on quality framework levels and standards achieved by other practices (e.g. comparative practice data, audits from other practices) or previous audits within the practice
- **Observing practice** You can collect information from: computer registers; medical records; questionnaires—patients, staff, or GPs; data collection sheets (e.g. drugs in doctor's bag are all in date)
- **Comparing results with standards** Consider why standards have not been met—what should be done? Who's going to do it? When? How?
- **Repeating the audit cycle** To ensure action taken is effective

Significant event review Process in which individual episodes (when there has been a significant occurrence either beneficial or deleterious) are analysed, in a systematic and detailed way to ascertain what can be learnt about the overall quality of care, and to indicate changes that might lead to future improvements. Methods of reporting—Table 2.1.

Table 2.1 Methods for reporting significant event audits

Reporting method 1	Reporting method 2
Description of event This should be brief and can be in note form *Learning outcome* This should describe the aspects which were of high standard and those that could be improved. Where appropriate, it should include why the event occurred *Action plan* The decision(s) taken need to be contained in the report. The reasons for these decisions should be described together with any other lessons learned from the discussion	*What happened?* *Why did it happen?* *Was insight demonstrated?* *Was change implemented?*

What is the difference between audit and research? Research is discovery of new knowledge. Research differs from audit as research aims to establish what best practice is globally; audit aims to discover how close practice is to best practice on a local level and identify ways of improving care.

Drug company research ➔ p. 122

The research process GPs may be involved in research at many levels—as part of an academic department, in a research general practice, or just by taking part in a project. If considering doing some research:

- Turn your idea into a specific research question What is your aim/hypothesis? Review the literature. Is your idea novel? Why does it matter?
- Design the study and develop your methods Involve participants/other researchers. Qualitative/quantitative methodology? Survey design/sample size? How will you choose/randomize participants? Who will do the work?
- Obtain permissions, funding, and Primary Care Research Network (PCRN) acceptance Ask the healthcare trust in which the research will be performed. Ensure that you contact the research governance officer of the organization within which your research will be carried out, and comply with local and national requirements. Obtain ethics permission. Obtain funding to cover the costs of the study and staff required. Gain acceptance onto the PCRN (or another research network) portfolio
- Collect and collate data Remember data protection (➔ p. 47)
- Analyse and interpret data Consider involving a professional statistician for quantitative data; think about the implications of the study findings; identify how findings can be put into practice
- Write up and disseminate findings Journal articles; conference presentations; press releases; submission for higher degrees

National Institute for Health Research (NIHR) Commissions and funds NHS and Social Care research in England, and provides infrastructure to support both studies and researchers within the NHS. 🖱 www.nihr.ac.uk

University departments of general practice Every UK medical school has a department of general practice. There are few GP academic posts but these departments are valuable sources of advice and support if you contemplate doing any original research of your own.

Ethics An ethics committee must pass all medical research involving human participants. Information, contacts, and application forms are available from the Health Research Authority. 🖱 www.hra.nhs.uk

Funding Numerous sources of funding for primary care research are available (including NIHR, RCGP, Medical Research Council, and Wellcome Trust) but all are keenly fought for. Contact your local medical school academic primary care department for advice and help if you are thinking of applying for research funding.

Further information

Healthcare Quality Improvement Partnership (2016) Best practice in clinical audit. 🖱https://www.hqip.org.uk/resource/best-practice-in-clinical-audit/

Outcomes in general practice

Within the NHS there are ↑ demands for accountability/improvements in quality of care. Measuring this requires use of appropriate outcomes.

Quality and Outcomes Framework ➔ p. 33

Patient satisfaction Implies meeting both the wants and the needs of the patient. Satisfaction measures are increasingly being used to judge the effectiveness of the NHS. Surveys of satisfaction show ~80% of patients are overall satisfied with GP care, but if questioned more specifically about different components of care (e.g. information provided, communication, etc.), fewer than half are completely satisfied.

Revalidation Measuring patient satisfaction of ≥34 patients, using a validated tool approved by the GMC, is a requirement for GPs once in every 5y revalidation cycle. GPs must reflect on results and address any development needs.

Peer feedback A tool widely used in business to assess worker performance. It may take many forms from team meetings during which colleague feedback is encouraged, through annual appraisal by a practice manager to 360° or multisource feedback forms.

Revalidation Feedback from ≥15 professional colleagues representing the range of your professional activities using a validated *multisource feedback* (MSF) tool approved by the GMC is a compulsory element of revalidation and must be performed once in every 5y cycle. GPs must reflect on results and address any development needs.

Comparative practice data The government's '*transparency agenda*' has resulted in publication of data held by government departments on the performance of public services including general practice. Data now in the public domain are listed in Box 2.3.

Box 2.3 Examples of GP practice data available in the public domain

- Local demography—age, ethnicity, deprivation
- Life expectancy of the practice population
- Overall QOF achievement
- QOF prevalence data, e.g. rheumatoid arthritis, DM, CVD
- Cancer care—new cancer cases, emergency cancer presentations, cancer prevalence based on QOF data, cancer screening uptake rates
- Child health—A&E attendances, emergency and elective admissions, outpatient attendances
- Antibiotic prescribing—prescribing rates, proportion of quinolones and broad-spectrum antibiotics
- Patient satisfaction
- Clinical staff details—names and roles
- CQC inspection reports
- GP income

Referral rates There are wide variations in referral rates (3–12/100 consultations) not accounted for by population characteristics. Experience in a specialty ↑ referrals implying high referrers are not always inadequate.

Referral management schemes Attempts have been made to judge appropriateness of referrals either at practice or at locality level through referral management schemes. All referrals are scrutinized to judge whether the referral is needed at all, and (if referral is warranted) whether referral is being made to the most cost-effective service.

Prescribing rates There are wide variations in prescribing rates, e.g. variation in the rate of statin prescription cannot be accounted for by population characteristics; prescription rates for antibiotics for minor illness vary widely between GPs. Whether and how these reflect quality of care are controversial but comparison of practice prescribing data within localities and prescribing quality targets are used to drive down prescribing costs.

Procedures Comparisons of procedure outcome (e.g. inadequate smear rates, diabetic outcome measures, immunization rates) between practices can be a way to identify individuals or practices clearly performing less well than others. The reasons must then be investigated.

Doctors' ability to detect illness There are wide variations between GPs in their ability to detect certain illnesses, e.g. mental illness. GPs adept at identifying mental health problems have: empathy, early eye contact, use directive rather than closed questioning; clarify the complaint at an early stage. Whether this is a marker of quality of care or just the diversity of general practice is debatable.

Compliance/concordance ➔ p. 116

Further information

National General Practice Profiles 🖱 https://fingertips.phe.org.uk/profile/general-practice
RCGP Colleague feedback. 🖱 www.rcgp.org.uk/training-exams/practice/revalidation/guide-to-supporting-information-for-appraisal-and-revalidation/feedback/colleague-feedback.aspx
RCGP Patient feedback. 🖱 www.rcgp.org.uk/training-exams/practice/revalidation/guide-to-supporting-information-for-appraisal-and-revalidation/feedback/patient-feedback.aspx

Chapter 3

Consulting with patients

The consultation *60*
Consultation models *62*
Telephone consulting and home visits *64*
Referrals and electronic media *66*
The doctor's bag *68*
Evidence-informed decision-making *70*
Guidelines, protocols, and integrated care pathways *72*
Breaking bad news *74*
Confirmation and certification of death *76*
Organ donation *78*

The consultation

Over the past 15y, in the UK, there has been a 40% ↑ in demand for GP appointments. Each patient has an average of 5.5 appointments/y. Older people have the highest consultation rate (those >80y have 13.5 appointments/y), and the consultation rate is set to ↑ as the population ages.

Potential barriers to effective communication Lack of time, language problems, differing gender, age, ethnic or social background of doctor and patient, 'sensitive' issues to address, 'hidden' or differing agendas, prior difficult meetings, and lack of trust between doctor/patient.

The consultation Good communication is essential for all aspects of a GP's work. The consultation is the cornerstone of general practice and focuses on successful information exchange. Various consultation models exist (➔ p. 62) to help GPs evaluate their consultations and make optimum use of the time available. There is no 'correct' way to perform a consultation. Approach will vary according to situation and participants.

Patient centredness Means that the patient's viewpoint is considered and integrated into the diagnosis and decision-making process (Figure 3.1). It improves patient satisfaction and may improve health outcomes. It consists of 6 interactive components:

- Exploring the disease and illness experience
- Understanding the whole person in context
- Finding common ground regarding management
- Incorporating prevention and health promotion

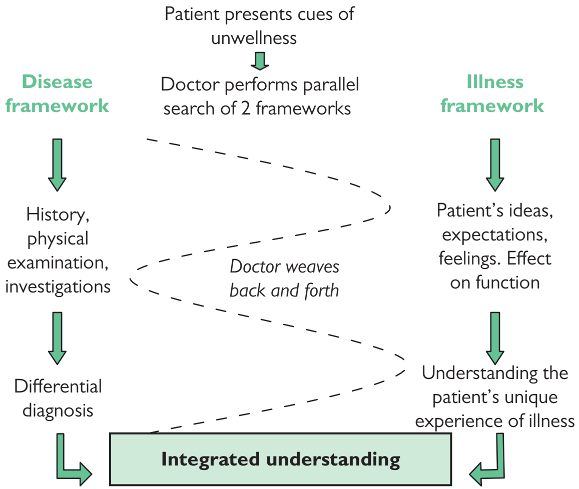

Patient presents cues of unwellness
Doctor performs parallel search of 2 frameworks
Disease framework
Illness framework
History, physical examination, investigations
Patient's ideas, expectations, feelings. Effect on function
Doctor weaves back and forth
Differential diagnosis
Understanding the patient's unique experience of illness
Integrated understanding

Figure 3.1 The patient-centred process

Reproduced with permission from Stewart, M. et al. *Patient-centred medicine: transforming the clinical method*. Sage Publications. Copyright © 1995.

- Enhancing the doctor–patient relationship
- Being realistic

Patient records ➔ p. 44

Patient recall Many studies suggest that >50% (some estimate up to 90%) of information has been forgotten within a few minutes of leaving the surgery. Characteristics of memorable information:

- The patient perceives it as important
- The patient understands it (avoid the use of jargon and medical terms, keep language brief and simple, support information with sketches/diagrams ± patient information sheets)
- The information is given early in the consultation
- The information is given in small chunks (not too much at once)

Consultation length Although consultations are usually booked at 10min intervals in the UK, average consultation length is now 11.7min. Instead of managing acute illness, GPs now focus on management of long-term conditions and multimorbidity, resulting in ↑ complexity of consultations. Despite this, UK consultations are still a third shorter than those in most other parts of the world, e.g. the USA, Switzerland, New Zealand, Belgium, and Australia.

Timekeeping Running late is stressful and frustrating for patients. General practice does not fit conveniently into 10min (or any other size) slots. Even the best time keepers occasionally run late. *Tips*:

- **Endeavour to run to time** Start on time; make appointments long enough (e.g. book double appointments for difficult problems, schedule catch-up slots in the middle of surgeries, change to longer appointments); break difficult problems or multiple problems up into chunks
- **If you are running late** Ask reception staff to apologize to patients as they check in, and tell them the expected delay

Benefits of longer consultation times Include:

- ↑ patient and doctor satisfaction
- ↑ detection of psychosocial problems
- Improved doctor–patient communication
- ↑ health promotion
- ↓ reconsultation rates
- ↓ minor illness prescribing

'Difficult' patients Characterized by: frequent presentation—the top 1% of attenders at GP surgeries generate 6% of GP workload; highly complex, often multiple problems—some explicable, others not; and exasperation generated between patient and doctor

❗ This is a two-way process. Some GPs report more difficult patients than others. The problem relates to the GP's perception of patients as well as the patients themselves.

Management strategy Do a detailed review of notes ± chart of life.

- Agree contacts (e.g. limit to 1 GP, agree appointment frequency)
- Agree an agenda within consultations, e.g. problem list—1 problem/visit
- Employ reattribution techniques—see Somatization disorder, ➔ p. 975
- Avoid unnecessary investigation and referral
- Be aware of your own reaction to the patient
- Acknowledge that such patients may be genuinely ill
- Consider psychiatric diagnoses—especially chronic anxiety, depression, somatization disorder. Screening questionnaires can be useful
- Consider referral for CBT and/or specialist mental health support

Consultation models

Consultation models are not rules; they provide a toolkit of different techniques for GPs to apply to their consultations. A brief overview of the most commonly used models is presented here—for more information, consult the original texts:

The medical model Traditional medical school model. History taking → examination → investigation → diagnosis → treatment → follow-up. Does not recognize the complexity/diversity of the consultation in general practice.

Balint, 1957 *The Doctor, His Patient and The Illness*—a philosophy rather than a consultation model:

- Psychological problems are often manifested physically
- Doctors have feelings. Those feelings have a role in the consultation
- Doctors need to be trained to be more sensitive to what is going on in the patient's mind during a consultation

Reference: Churchill Livingstone; ISBN: 0443064601.

Berne, 1964 *Games People Play*—describes how to recognize behaviours ('games') patients might use and roles patient and doctor might adopt—'Parent, Adult and Child'.

Reference: Penguin Books; ISBN: 0140027688.

RCGP, 1972 *The triaxial approach*—physical, psychological, and social aspects of the consultation.

Reference: Working party of the RCGP, 1972.

Becker and Maiman, 1975 *Health Belief Model*—Involves exploration of ICE—Ideas, Concerns, and Expectations of the patient. 5 elements:

- Health motivation
- Perceived vulnerability
- Perceived seriousness
- Perceived costs/benefits of an action
- Cues to action—stimuli/triggers for beliefs

Reference: *Med Care* 1975;**13**:10–24.

Byrne and Long, 1976 *Doctors talking to patients*—6 aspects:

1. Doctor establishes a relationship with the patient
2. Doctor attempts to/actually discovers the reason for attendance
3. Doctor conducts verbal ± physical examination
4. Doctor, or doctor + patient, or patient consider the condition
5. Doctor (or occasionally the patient) details treatment and investigation
6. Consultation is terminated—usually by the doctor

Reference: RCGP; ISBN: 0850840929.

Stott and Davis, 1979 *Exceptional potential of the consultation*. 4 tasks:

1. Management of presenting problems
2. Management of continuing problems
3. Modification of help-seeking behaviour
4. Opportunistic health promotion

Reference: *JRCGP* 1979;**29**:201–5.

Helman's folk model, 1981 *Disease vs illness in general practice*:

- What has happened?
- Why has it happened?
- What would happen if nothing were done about it?
- What should I do and who should I consult for further help?
- Why to me?
- Why now?

Reference: *JRCGP* 1981;**31**:548–52.

Pendleton *et al.*, 1984 *The doctor's tasks*:

- Define the reason for patient's attendance
- Consider other problems—continuing problems and risk factors
- Choose an appropriate action for each problem—involves negotiation between doctor and patient
- Achieve a shared understanding of the problem between doctor/patient
- Involve the patient in the management and encourage the patient to accept appropriate responsibility
- Use time and resources appropriately
- Establish and maintain a relationship between doctor and patient

Reference: Oxford University Press; ISBN: 0192632884.

Neighbour, 1987 *The Inner Consultation*. Checkpoints:

- Connecting—doctor establishes rapport with the patient
- Summarizing—doctor clarifies the patient's reason for consulting
- Handing over—doctor/patient negotiate and agree a management plan
- Safety netting—planning for the unexpected; managing uncertainty
- Housekeeping—doctor is aware of his/her own emotions

Reference: Petroc Press; ISBN: 1900603675.

Fraser, 1992 and 1999 *Areas of competence*:

1. Interviewing and history taking
2. Physical examination
3. Diagnosis and problem-solving
4. Patient management
5. Relating to patients
6. Anticipatory care
7. Record-keeping

Reference: Butterworth Heinemann; ISBN: 0750640057.

Kurtz and Silverman, 1996 and 2002 *Calgary—Cambridge Observation Guide*. 5 tasks:

1. Initiating the session
2. Gathering information
3. Building the relationship
4. Giving information—explaining and planning
5. Closing the session

Reference: *Med Educ* 1996;**30**:83–9 and *Acad Med* 2003;**78**(8):802–9

Warren, 2002 4 avenues of consultation analysis (BARD):

- Behaviour—non-verbal and verbal—needs of patient/personality of GP
- Aims—purpose of the consultation and priorities
- Room—setting for the consultation
- Dialogue—tone of voice, what is said, etc.

Reference: *Update* 2002;**5 Sept**:152–4.

Further information

Bradford VTS. Consultation models. 🖱 www.bradfordvts.co.uk

Telephone consulting and home visits

To cope with ↑ demand for GP appointments, the past 20y have seen the proportion of GP consultations taking place over the telephone ↑ from 3% to 12%; simultaneously, the proportion of consultations taking place in patients' own homes has ↓ from 9% to 3.5%.

Emergency telephone consultations Nearly all requests for emergency care are made by telephone. *General rules*:

- Train surgery staff to handle distressed callers, recognize serious problems, and act appropriately when such calls are received
- Appear helpful from the outset. Keep calm and friendly—even if provoked; worried callers often appear abrupt or demanding
- Record the time of the call, date, patient's name, address the patient is at and a contact telephone number, brief details of the problem, and action taken (even if calls are being recorded)
- Collect only information you need to decide whether a visit/urgent surgery appointment is necessary. If a visit is necessary, collect enough information to decide how quickly the patient should be seen and whether extra equipment or help is needed. If a visit is not necessary, decide whether other actions, such as an urgent surgery appointment, are required
- If giving advice, make it simple and in language the patient can understand. Repeat to make sure it has been understood. Consider asking patients/carers to repeat what you have told them. Always tell callers to ring back if symptoms change or they have further worries
- If a visit is indicated, ensure the address is correct and ask for directions if you are not sure where to go. Try to give a rough arrival time
- In some cases (e.g. major trauma, large GI bleeds, MI, stroke, burns, overdoses), call for an emergency ambulance at once
- If a call seems inappropriate, consider the reason for it, e.g. depression might provoke recurrent calls for minor ailments

⚠ If in doubt—see the patient.

Routine telephone consultations May take various different forms:

- Telephone triage systems to filter requests for surgery appointments
- Telephone clinics where patients are free to call with their problems
- Telephone message books
- Bookable telephone slots in surgery time

The telephone is a useful way to answer simple queries without wasting surgery time. Examples include:

- Consultations for minor, self-limiting conditions or conditions not requiring an examination
- Follow-up of surgery consultations, e.g. to give results, or offer management advice or a prescription following investigations

The biggest drawbacks of telephone consultations are:

- Inability to examine the patient
- Lack of visual cues to aid communication—be alert for verbal cues (e.g. lowering of the voice, hesitations, signs of distress). Ask about ideas/concerns, and invite the patient to ask questions

❗ Before giving advice, ensure you have sufficient information upon which to base your judgement. If examination is needed, see the patient.

Remote video consultations A large number of both private and NHS GP services now routinely use video consultation platforms (such as Skype) to deliver GP consultations. Feedback from both patients and healthcare practitioners has been positive. From the GP perspective, consultations take place in much the same way as standard telephone consultations but the doctor is able to see the patient, can perform rudimentary visual examination, and is also able to pick up visual cues from the patient.

Home visiting Home visits may be routine checks for housebound patients or emergency visits for patients temporarily unable to get to the surgery. Home visits done in working hours are usually done by practices under their GMS/PMS contract but, in some areas, home-visiting services are provided by the PCO and practices are able to 'opt out'.

Routine visits Conducted like ordinary surgery consultations. Seeing patients in their own home may give valuable extra information.

Emergency visits

- Try to stick to the problem you have been called about; take a concise history and examine as appropriate
- Make a decision on management, and explain it to the patient and any carers in clear and concise terms they can understand. Repeat advice several times ± write it down
- Record history, examination, management suggested, and advice given
- Always invite the patient/carers to call you again should symptoms change, the situation deteriorate, or further worries arise
- For inappropriate calls, take time to educate the patient and/or carers about self-management and use of emergency GP visiting services; always consider hidden reasons for seemingly unnecessary visits

Being prepared

- Ensure you have a reliable car with a full tank of fuel
- Have a good street map and in-car electronic navigation system
- Carry a mobile phone and large, strong torch in the car
- Check your drug box is fully stocked and all items are in date
- Check all equipment carried is operational and carry spare batteries
- Carry a list of emergency telephone numbers and know which pharmacies have extended opening hours

Safety and security

- In all cases, ensure someone else knows where you are going, when to expect you back, and what to do if you do not return on time
- If going to a call you are worried about, either take someone with you to sit in the car or call the police to meet you there before going in
- If you are uncomfortable, make sure you can get out. Note the layout of the property, and make sure you have a clear exit route to the door
- Set up your mobile phone to call the police or your base at a single touch. Consider carrying an attack alarm
- If possible, have separate bags for drugs and consultation equipment; leave the drug box locked out of sight in the boot of the car when visiting

Referrals and electronic media

Referral letters Good communication is essential when referring patients to other doctors/agencies. Electronic pre-filled word processing templates may add some information automatically, but ensure all referral letters include:

- Address of the referrer (including telephone number, if possible), name and address of registered GP if not the referrer, and date of referral
- Name, address, telephone number, and date of birth of the patient (and any other identifiers available, e.g. hospital or NHS number)
- Name of the person to whom the patient is being referred (or department if not a named individual)
- Presenting condition—history, examination, investigations already performed with results, treatments already tried with outcomes
- Relevant past medical history and family history
- Current medication, and any intolerances/allergies known
- Reason for referral (what you want the recipient of the letter to do), e.g. to investigate symptoms, to reassure parents
- Any other relevant information, e.g. social circumstances
- Signature and/or name in legible format of referrer + GMC/Nursing and Midwifery Council number
- Ensure a copy of the referral is stored on the patient's electronic record

Referral rates ➔ p. 57

Referral management systems ➔ p. 57

NHS e-Referral Service (eRS) Since October 2018, Trusts will only accept e-referrals from GPs. Once a referral is made, patients can book an appointment at a time and venue of their choice either in the GP surgery at the point of referral, or later on the telephone or online.

Use of e-mail in the GP surgery

Dissemination of information E-mail is widely used in the UK to disseminate (cascade) information to GPs, e.g. NICE, DH, MHRA, PCOs.

Communication with doctors/other healthcare providers Many communications between doctors occur by e-mail both within practices and also between practices/other healthcare providers (e.g. consultant advice by email). ❗ E-mail transfer between non-NHS e-mail accounts should never be considered secure or confidential.

Communication with patients ~80% of the UK population now has access to e-mail. Cyber-savvy patients increasingly want to be able to communicate with healthcare professionals by e-mail, but it has been used relatively little for communication with patients due to concerns over quality of e-mail content, time lag, confidentiality, and liability. The NHS eConsult system is likely to increase electronic communication between GPs and patients. The eConsult system uses standard questionnaires and templates to exclude urgent conditions unsuitable for eConsult and collect important information for the consulting GP.

Guidelines for e-mail consultations with patients

- Establish turnaround time; do not use e-mail for urgent matters
- Warn users that e-mail is not secure and that they cannot assume confidentiality just because they are communicating with a doctor; advise patients not to use a work or multi-user e-mail account
- Retain copies of e-mail communications with patients

- Instruct patients to put their name and date of birth in the message, and the category of transaction in the subject line of the message for filtering: e.g. prescription, appointment, medical advice
- Configure an automatic reply to acknowledge receipt of messages
- Send a new message to inform the patient of completion of the request; ensure that others are not copied into the reply
- Avoid giving person-specific information or confirming information given by the patient—it may not be the patient
- Never write anything that you would not be happy to see printed on a newspaper's front page
- Append a standard block of text to the end of messages which contains the GP's name, contact information, and reminders about security and the importance of contacting the practice by telephone for emergencies
- Explain to patients that their messages should be concise
- Remind patients when they do not adhere to the guidelines

Social media Internet-based websites/tools allowing users to create/share content between networks of people, e.g. Facebook, LinkedIn, Twitter, YouTube. Social media are used successfully by doctors to:

- Establish wider/more diverse social and professional networks
- Engage the public and colleagues in debates
- Facilitate public access to accurate health information
- Improve patient access to services

Risks of social media use

- Loss of personal privacy
- Potential breaches of confidentiality
- Online behaviour that might be perceived as unprofessional, offensive, or inappropriate by others
- Risks of posts being reported by the media or sent to employers

Guidance on social media use The BMA, GMC, and RCGP have all produced guidelines for doctors on the use of social media. *Key points*:

- Social media blurs boundaries between public/professional lives
- Adopt conservative privacy settings where available, but be aware that not all information can be protected on the web
- Ethical and legal duties to protect patient confidentiality apply equally on the Internet as to other media
- It is inappropriate to post informal, personal, or derogatory comments about patients or colleagues on public Internet forums
- Doctors who post online should declare any conflicts of interest
- Do not accept Facebook friend requests from current/former patients
- Defamation law can apply to any comments posted on the web made in either a personal or professional capacity
- Be conscious of online image and its impact on professional standing

Further information

BMA Social media guidance for doctors. 🖱 www.bma.org.uk
GMC Doctor's use of social media. 🖱 www.gmc-uk.org
RCGP Social media highway code. 🖱 www.rcgp.org.uk/social-media

The doctor’s bag

⚠ The GP’s bag must be lockable and not be left unattended during home visits. If left in the car, keep the bag locked and out of sight—preferably in the boot. Consider having a separate bag for drugs and consultation equipment and only get the drug bag out of the boot of the car if it is needed. Keep the bag away from extremes of temperature.

Consider including the following in your doctor’s bag. Exact contents will vary according to location and circumstances:

Diagnostic equipment

- Stethoscope
- Sphygmomanometer
- Thermometer
- Gloves, lubricating jelly, and tissues
- Torch
- Otoscope
- Ophthalmoscope
- Tongue depressors
- Peak flow meter
- Pulse oximeter
- Fluorescein sticks
- Urine dipsticks
- Capillary blood glucose tester and appropriate test strips
- Tourniquet, vacutainer (or syringe), and needles
- Patella hammer
- Swabs
- Specimen containers
- Vaginal speculum ± sponge forceps
- Fetal stethoscope/Doppler

Administrative equipment

- Mobile telephone ± charger
- Laptop computer with access to full patient record
- Prescription pad
- List of useful telephone numbers
- BNF/MIMS
- Envelopes/notepaper
- Electronic navigation aid ± map
- Obstetric calculator
- Peak flow chart/wheel
- Note book
- Temporary resident records
- List of local chemists and extended opening times
- Small amount of change for parking, etc.
- Pathology/X-ray forms (if electronic forms are not used)
- Controlled drugs record book (if controlled drugs are carried)
- Quick reference text, e.g. Oxford Handbook of General Practice

Other equipment

- Airway ± Laerdal mask
- Oxygen cylinder and mask with reservoir bag
- Automated external defibrillator
- Nebulizer
- Spacer device
- IV cannula
- IV giving set and fluids
- Needles/syringes
- Bandages
- Gauze swabs
- Adhesive plasters
- Scissors
- Skin closure strips
- Suturing equipment/skin glue
- Urinary catheter and bag
- Antiseptic sachets
- Dressing pack
- Sharps box

Drugs for the doctor's bag *Consider*:

Injectables

- Adrenaline (epinephrine)
- Naloxone
- Benzylpenicillin injection
- Cefotaxime injection
- Lorazepam/diazepam
- NSAID, e.g. diclofenac
- Local anaesthetic, e.g. lidocaine
- Opioid analgesic, e.g. morphine, oxycodone
- Antiemetic, e.g. domperidone, prochlorperazine
- Antihistamine, e.g. chlorphenamine
- Hydrocortisone injection
- Diuretic, e.g. furosemide
- Syntometrine®
- Glucagon ± IV glucose
- Major tranquillizer, e.g. haloperidol, chlorpromazine
- Thrombolytic therapy (if >½h from nearest acute hospital and trained)

Oral drugs

- Antacid
- Antibiotics (adult tablets and paediatric sachets), e.g. amoxicillin + erythromycin/clarithromycin + trimethoprim/nitrofurantoin
- Antihistamine
- Rehydration tablets/sachets
- Aspirin
- Lorazepam
- Paracetamol tablets + suspension
- Prednisolone tablets (soluble)
- NSAID, e.g. ibuprofen

Other drugs

- GTN spray
- Bronchodilator for nebulizer
- Salbutamol inhaler + spacer
- GlucoGel® glucose gel
- Glycerol suppositories
- Rectal diazepam
- Diclofenac suppositories
- Domperidone suppositories

Drugs administered from a doctor's bag Should be in a suitable container and properly labelled with:

- Patient's name
- Drug name
- Drug dosage
- Quantity of tablets
- Instructions for use
- Relevant warnings
- Name and address of the prescriber
- Date
- Warning 'Keep out of reach of children'

⚠ Check drugs at least 2×/y to see they are still in date and usable; commercial databases (e.g. 🖱 www.doctorsbaguk.com) that store data about drugs in the GP's bag can be useful to alert you when drugs go out of date.

⚠ Record origin, batch number, and expiry date of *all* drugs administered to patients or dispensed to them to take themselves.

Further information

Drugs and Therapeutics Bulletin:

- Drugs for the doctor's bag 1—adults (May 2015)
- Drugs for the doctor's bag 2—children (June 2015)

Evidence-informed decision-making

Evidence-based medicine (EBM) is *'the conscientious, explicit and judicious use of current best evidence in making decisions about the care of the individual patient. It means integrating individual clinical expertise with the best available external clinical evidence from systematic research.'* (Sackett et al., 2018)

The 5 steps of EBM

1. Convert clinical information needs into answerable questions
2. Track down the best evidence with which to answer them (Box 3.1)
3. Critically appraise that evidence for its validity/usefulness
4. Apply the results of this appraisal in clinical practice
5. Evaluate your clinical performance, e.g. through audit (➔ p. 54)

Box 3.1 Useful sources of best evidence for GPs

- **NICE** (➔ p. 25) 🖱 www.nice.org.uk
- **Cochrane** Database of high-quality systematic reviews to inform healthcare decision-making 🖱 www.cochranelibrary.com
- **PubMed Central (PMC)** Free to use, searchable database of biomedical and life sciences journal literature 🖱 www.ncbi.nlm.nih.gov/pmc
- **Google Scholar** Freely accessible web search engine that indexes scholarly literature across an array of publishing formats and disciplines

Critical appraisal is the process of assessing and interpreting evidence by systematically considering its validity, results, and relevance. It is essential to avoid misinterpretation and misuse of evidence in practice.

Table 3.1 explains grading of evidence from most to least reliable; Table 3.2 is a glossary of common EBM/critical appraisal terms. Critical Appraisal Skills Programme (CASP) checklists are available to download from 🖱 www.casp-uk.net. Before integrating evidence into practice consider:

- Are the results of the study valid and applicable to my patient?
- What are the results?
- Will they help me in caring for my patients?

Table 3.1 Classification and grading of evidence: most → least reliable

Grade	Evidence level	Definition: evidence obtained from …
A	Ia	Meta-analysis of randomized controlled trials
	Ib	At least 1 randomized controlled trial
B	IIa	At least 1 well-designed controlled study without randomization, e.g. case–controlled study; cohort study
	IIb	At least 1 other type of well-designed quasi-experimental study
	III	Well-designed, non-experimental descriptive studies, such as comparative studies, correlation studies, and case studies
C	IV	Expert committee reports or opinions and/or clinical experience of respected authorities

Table 3.2 Glossary of terms used in EBM and critical appraisal

Term	Explanation
Systematic review and meta-analysis	Appraisal and synthesis of individual study reports using a rigorous and reproducible methodology for searching for and selecting studies included, thereby minimizing bias. Meta-analysis is systemic review using quantitative methods to summarize results
Randomized controlled trial (RCT)	Subjects are randomly assigned to an intervention or control group (no intervention, placebo or alternative intervention). Groups are compared to detect any differences in outcome
Case–control study	Identifies a group with an outcome of interest (cases) and another without (controls), and looks for history of a particular exposure
Cohort study	Identifies 2 groups (cohorts) of patients: 1 that received the exposure of interest, and 1 that did not. Then follows the cohorts forward looking for the outcome of interest
Cross-sectional study	Observation of a population at a set time or over a defined period of time
Cost–benefit analysis	Assesses whether cost of an intervention is worth the benefit by measuring both in the same (usually monetary) units
Bias	Can take many forms but describes any systematic errors that may distort outcome or interpretation of trial evidence
Confidence interval (CI)	Quantifies uncertainty. Usually reported as 95% CI, which is the range of values between which it is 95% sure that the true value lies
Control and experimental event rates	Rate at which events occur in a control group (CER) or experimental group (EER). CER/EER = event rate ÷ total number in the group, e.g. if 10 out of 100 have an event in the control group, CER = 0.1. May be expressed as a percentage (e.g. 10%) or proportion (e.g. 0.1)
Absolute risk ↑ or ↓	Absolute arithmetic difference in rates of outcomes between experimental and control participants in a trial (= CER − EER)
Relative risk or risk ratio (RR)	Ratio of risk in the treated group (EER) to risk in the control group (CER). RR = EER/CER. Used in RCTs and cohort studies. If RR = 1 there is no difference between the two groups for that measure
Relative risk ↓ (RRR)	Difference between the EER and CER (EER − CER) divided by the CER. Usually expressed as a percentage
Number needed to treat or harm (NNT/NNH)	Measure of the difference between active intervention and control in terms of benefit or harm. Equal to 1 ÷ absolute risk reduction (or increase). For NNT, an NNT of 1 indicates 100% effectiveness; the closer the NNT is to 1, the more effective the intervention. For NNH, the higher the value, the safer the intervention
Odds ratio (OR)	Odds of an event are the number of events divided by the number of non-events. OR is the ratio of the odds in the experimental divided by the odds in the control group. Often expressed as a percentage
Negative predictive value ➔ p. 145	*Sensitivity* ➔ p. 145
Positive predictive value ➔ p. 145	*Specificity* ➔ p. 145

Further information

Sackett SE, **et al.** (2018). *Evidence-Based Medicine: How to Practice and Teach EBM*, 5th edition. Elsevier: Churchill Livingstone.

Guidelines, protocols, and integrated care pathways

Clinical guidelines Defined as '*user-friendly statements that bring together the best external evidence and other knowledge necessary for decision making about a specific health problem*'. Over recent years there has been a dramatic ↑ in publication of guidelines and protocols. They aim to ↓ harmful or expensive variations in clinical practice, improve healthcare outcomes and encourage rapid dissemination of useful innovations. Good clinical guidelines have 3 properties—they:

- Define practice questions and identify all their decision options and outcomes
- Identify, appraise, and summarize best evidence about prevention, diagnosis, prognosis, therapy, harm, and cost-effectiveness
- Identify the decision points at which the evidence needs to be integrated with individual clinical experience and clinical circumstances in deciding a course of action

Advantages and disadvantages of guidelines

Advantages

- Provide guidance for busy clinicians—a consistent basis for decision-making
- Practical framework for common problems and chronic disease
- Summarize the available research evidence
- Can be used as a basis for continuing medical education
- Justification for expenditure—can aid cost-effective use of limited resources
- Facilitate the audit cycle

Disadvantages

- Poor quality guidelines can reinforce poor practice
- Lack of relevance of the guidelines to the clinical setting—much of the 'evidence' used to develop guidelines comes from secondary care and may not reflect the situation in primary care
- Tendency to uniformity—can stifle innovation
- Resistance to change—new methods may not be considered until a new guideline is produced
- Increased risk of litigation
- Cost—guidelines are time consuming to develop and update
- Lack of ownership—guidelines developed by others may not feel relevant
- Difficulties in implementation—guidelines that are not user-friendly and well disseminated will not be used

Before starting to use a guideline Always ask:

> '*Is this guideline valid, important and applicable in my practice?*'

If the answer is yes then consider:

- What barriers exist to implementation?
- Can they be overcome?
- Can you enlist collaboration of key colleagues?
- Can you meet the educational and administrative conditions that are likely to determine the success or failure of implementing the strategy?

Protocol The term reserved for guidelines at the more rigid end of the spectrum. These are very specific guidelines which are expected to be followed in detail, with little scope for variation, e.g. resuscitation protocols.

Integrated care pathway (ICP) ICPs amalgamate all the anticipated elements of care and treatment of the multidisciplinary team, for a particular patient group in order to achieve agreed outcomes. Any deviation from the plan is documented as variance—the analysis of which provides information for the review of current practice. ICPs aim to:

- Facilitate introduction of guidelines and systemic audit into clinical practice
- Improve multidisciplinary communication and care planning
- Reach or exceed existing standards
- Decrease unwanted practice variation
- Improve clinician–patient communication and patient satisfaction
- Identify research and development questions
- Cross the interface between primary, secondary, and social care

Grading/classification of evidence Table 3.1, ➔ p. 70

Further information

eGuidelines (free registration required) 🖱 www.eguidelines.co.uk
National Guidelines Clearing House (US) 🖱 www.guideline.gov
NICE 🖱 www.nice.org.uk
Scottish Intercollegiate Guidelines Network 🖱 www.sign.ac.uk

Breaking bad news

GPs break bad news frequently, but it is never easy.

Why is breaking bad news hard?

- **Admission of failure** When we tell patients bad news, it is often an admission that we have failed. When we fail we naturally question what we have done and when looking at our practice in retrospect, it is easy to find fault. Feelings of guilt are common
- **Fear of the reaction of the patient** We all have a desire to avoid unpleasantness but sharing information with patients may be a positive way forwards. Even if news is bad, it gives patients control of the situation

Guidelines for sharing bad news with a patient

DO

- Plan the consultation as far as possible. Check the facts first, and ensure you have all the information. Ensure privacy and freedom from interruption
- Set aside enough time
- Ask if the patient would like a relative or friend with them. Make sure you introduce yourself and find out their name and relationship to the patient
- Make eye contact—watch for non-verbal messages. Sit at the same level as the patient
- Use simple and straightforward language
- Allow silence, tears, or anger
- Be prepared to go over facts again
- Answer questions
- Reflect on what the patient or relative has said to allow you to modify your understanding of their feelings
- Take into account the patient's current health, e.g. if in pain, then sort out the pain and schedule a further discussion when the patient is more comfortable
- Offer ongoing support

DON'T

- Lie or fudge the issue
- Get your facts wrong
- Break bad news in public
- Give the impression of being rushed or distant
- Give too much information. It is better to be concise—the finer points can be filled in later
- Interrupt or argue
- Say that 'nothing can be done'—there is always something that can be done
- Meet anger with anger
- Say you 'know how they feel'—you don't
- Be frightened to admit you don't know something
- Use medical jargon
- Leave the patient with no follow-on contact
- Agree to withhold information from the patient

Common problems

- What if the relatives do not want you to tell the patient? With adults of sound mind, information is confidential to the patient and can only be released, even to close relatives, with the patient's permission. Relatives who say they do not want the patient to know often do so to protect their relative. It is important to recognize they know your patient best. First, explore their worries and point out the difficulties of the patient not knowing. Often once a relative realizes that the patient knows things are not right and needs help and support to face the situation, they come round to the patient being told. Stress that you will not lie to a patient if asked directly
- How do you know if the patient wants to know? Most people (80–90%) *do* want to know. Assume this is the case and then feel your way carefully. Give the patient ample opportunity to say that they do not want to know
- How do you respond to questions you cannot answer? The best way to deal with this is to say that you do not have all the answers but will answer when you can, find out what you can, and say when you do not know

Bereavement, grief, and coping with loss ➔ p. 102

Confirmation and certification of death

English law *does not* require a doctor:

- To confirm death has occurred or that 'life is extinct'. A doctor is only required to certify what, in their opinion, was the cause
- To view the body of a deceased person. There is no obligation to see/examine a body before issuing a death certificate
- To report the fact that death has occurred

English law *does* require the doctor who attended the deceased during the last illness to issue a certificate detailing the cause of death. Certificates are provided by the local Registrar of births, marriages, and deaths. A special certificate is needed for infants of <28d old.

Death in the community 1 in 4 deaths occur at home.

Expected deaths In all cases, advise relatives to contact the undertakers; if you are not the patient's usual GP, ensure he/she is notified.

- **Patient's home** Visit as soon as is practicable
- **Residential/nursing home** If possible, the GP who attended during the patient's last illness should visit and issue a death certificate. The 'on-call' GP is often requested to visit. There is no statutory duty to do this but it is reassuring for the staff at the home and often necessary before staff are allowed to ask for the body to be removed

Unexpected and/or 'sudden' death If called, advise the attendant to call the emergency services.

Cremation Before a person can be cremated, Cremation Regulations (2008) require 2 doctors to complete a certificate to establish identity and that the cause of death is not suspicious. The person arranging the funeral may see the forms and pays a fee to each doctor. 2 parts:

- **Cremation 4** Completed by the patient's usual medical attendant—in the community, usually his/her GP
- **Cremation 5** Completed by another doctor who must have held full GMC registration (or equivalent) for ≥5y and is not connected with the patient in any way nor directly connected with the doctor who issued cremation form 4—usually a GP from another practice

⚠ Pacemakers and radioactive implants must be removed from the deceased before cremation can take place.

Notification of death to the coroner The coroner can be contacted via the local police but electronic notifications are preferred. Reporting a death to the coroner does not automatically entail a postmortem. Once circumstances are clear, the coroner may advise the GP to tick and initial box A on the back of the death certificate, which advises the registrar that no inquest is necessary. Deaths that must be reported to the coroner in England, Wales, and Northern Ireland are listed in Box 3.2.

❗ In Scotland, deaths are reported to a procurator fiscal. In addition to the list in Box 3.2, deaths of foster children and newborns must be reported.

Medical examiners From April 2019, local 'medical examiners' will start checking all death certificates issued by treating doctors for accuracy and coroner notification obligations. This effectively extends to burials the second certification function that already occurs for cremations. Initially medical examiners will only scrutinize deaths in hospitals (46% of deaths), but the scheme will eventually be extended to deaths in the community as well.

Recording deaths Death registers are useful. Routine communication of deaths to all members of the primary healthcare team and other agencies involved with the care of that patient (e.g. hospital consultants, social services) avoids the embarrassing and distressing situation of ongoing appointments and contacts being made for that patient. Record the death in the notes of any relatives/partner registered with the practice.

Bereavement, grief, and coping with loss ➔ p. 102

Benefits available after a death

- For widows/widowers ➔ p. 102
- Budgeting loans for funeral payments ➔ p. 105

Box 3.2 Deaths must be reported to the coroner if:

- The deceased was not seen by the certifying doctor *either* after death *or* within 14 days before death
- The death was violent or suspicious
- Death may be due to an accident (whenever it occurred)
- Death may be due to self-neglect or neglect by others
- Death may be due to an industrial disease or related to employment
- The death may be due to an abortion
- The death may be a suicide
- Death occurred during or shortly after police/prison custody
- Death occurred while the deceased was subject to compulsory detention under the Mental Health Act or a Deprivation of Liberty Safeguards authorization (DOLS)
- Cause of death is unknown, suspected to be unnatural, or there are any other concerning features

Patient advice

- What to do after someone dies 🖱 www.gov.uk/after-a-death
- What to do after a death in Scotland 🖱 https://www.gov.scot/Publications/2016/11/6948

Further information

Ministry of Justice (2012, updated 2018) Medical practitioners: guidance on completing cremation forms. 🖱 https://www.gov.uk/government/publications/medical-practitioners-guidance-on-completing-cremation-forms

Office for National Statistics (2010). Guidance for doctors completing medical certificates of cause of death in England and Wales. 🖱 https://www.gro.gov.uk/Images/medcert_July_2010.pdf#

Wessex LMCs Deprivation of Liberty Safeguarding (DOLS) and death. 🖱 https://www.wessexlmcs.com/deprivationoflibertysafeguardingdols

Organ donation

Over 6000 people in the UK are waiting for an organ transplant that could save or dramatically improve their life, but ~5000 transplants are carried out each year. There is a need for more donors. In 2017, >400 people died while awaiting a transplant.

Absolute contraindications to any organ donation:

- Creutzfeldt–Jakob disease (or other neurodegenerative diseases associated with infectious agents)
- Ebola virus infection
- Cancer that has spread within the past 12mo
- HIV or hepatitis C infection—although, rarely, organs may be used by other patients already infected

Donor cards and the NHS Organ Donor Register Potential donors should always discuss their wishes with their relatives. They can register their desire to donate their organs after death by adding their names to the NHS Organ Donor Register and obtaining an Organ Donor Card. Contact the NHS Organ Donor Line ☎ 0300 123 2323 or sign up online at 🖱 www.organdonation.nhs.uk.

❗ In Wales, unless an individual 'opts out', it is assumed that they have no objections to organ donation. It is anticipated that a similar 'opt-out' system will become operational in England in April 2020, and Scotland also plans to move to an 'opt-out' rather than 'opt-in' system in the future.

Blood donation New donors aged 17–65y are accepted. Donors may donate whole blood or platelets. Different blood transfusion services operate in the devolved nations of the UK. *Contacts*:

- England ☎ 0300 123 2323 🖱 www.blood.co.uk
- Northern Ireland ☎ 08085 534 666 🖱 www.nibts.hscni.net
- Scotland ☎ 0345 90 90 999 🖱 www.scotblood.co.uk
- Wales ☎ 0800 25 22 66 🖱 www.welsh-blood.org.uk

Bone marrow donation Bone marrow donation is open to people aged 18–49y who are blood donors (although may register at the time of first donation). Volunteers are HLA tissue-typed using DNA from white blood cells. Tissue type is recorded in the British Bone Marrow Registry. Donation involves a small operation in which bone marrow is harvested—usually from iliac crests.

Stem cell donation Peripheral stem cell donation is open to people aged 18–49y who are blood donors (although may register at the time of first donation). As for bone marrow donation, volunteers are HLA tissue-typed using DNA from white blood cells. Their tissue type is recorded within the British Bone Marrow Registry.

Donating stem cells involves daily injection of a growth factor (filgrastim) for 4d; this releases stem cells into the peripheral blood. Stem cells are harvested on day 5 using a cell separator from venous blood collected from 1 arm; the blood is returned through a vein in the other arm.

Cord blood stem cell donation When delivering their babies in certain hospitals in England, women can opt to donate cord blood for stem cell harvesting. *Further information*: 🖱 www.nhsbt.nhs.uk/cordblood

Surgical organ donation from living donors Two main types:

- **Donation at the time of routine operation** Femoral heads can be donated at the time of hip replacement; skin can be donated at the time of cosmetic surgery (e.g. apronectomy); amniotic membrane can be donated at the time of Caesarean section
- **Surgery to remove living organs** 1 kidney, part of lung, liver, or small intestine. Usually close relatives. Removal of the organ/part-organ involves a major operation for the donor. Risks to donor must be weighed against benefits to recipient

Heart-beating donation after death Donors must be maintained on a life-support machine at the time of death and until the organs are removed. The role of the GP in these situations is pre-emptive (information about Organ Donor Register) and to support families to make the decision whether to donate. Organs that can be donated: kidneys, heart, liver, lungs, pancreas, corneas, heart valves, bone, and skin.

Non-heart beating donation after death Donation can occur up to 24h after death (sometimes up to 48h). Tissues that can be donated:

No upper age limit	<60y
• Corneas	• Heart valves
• Skin	• Tendons
• Bone	

Donation of whole body for medical education The donor must give authorization for donation prior to death. Relatives should contact the medical school with which the donor has made arrangements after the donor's death. Medical schools arrange collection of the body and a simple funeral. Not all bodies are accepted. *Further information*: 🖱 https://www.hta.gov.uk/donating-your-body

Tissue or brain donation after death for research purposes Can be done in addition to donation for transplantation—organs for transplant are taken first. *Further information*: 🖱 https://www.hta.gov.uk/guidance-public/donating-your-tissue and 🖱 https://www.hta.gov.uk/guidance-public/brain-donation

Approach to relatives Many families find the act of donation a source of comfort. Even when the person who has died is on the Organ Donor Register, donation will be discussed with family members. However, if a person has agreed to donation prior to death and this is recorded on the Organ Donor Register, the family has no legal right to overrule that decision.

The coroner For any patient normally referred to the coroner, the coroner's permission must be gained before tissues are removed.

Further information

NHS Blood and Transplant ☎ 0300 123 2323 🖱 www.organdonation.nhs.uk

Chapter 4

Social aspects of primary care

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Social factors and health

> '*The task of medicine is to promote health, to prevent disease and to treat the sick . . . These are highly social functions*'

H.E. Sigerist, *Civilization and Disease* (1943)

Health inequalities are differences in people's health across the population and between specific population groups. They are socially determined by circumstances generally beyond an individual's control and unjust as they are avoidable if social inequalities were addressed.

Inverse care law Julian Tudor Hart's inverse care law states that '*the availability of good medical care tends to vary inversely with the need of the population served*'. This paradox is true across different diseases and healthcare systems. Health inequalities are not inevitable and addressing social factors can contribute significantly in reducing them; 60% of health improvement in the past century is not attributable to advances in medical care but instead to changes in social factors, such as better housing.

Deprivation Social deprivation is linearly associated with death from all causes, with no threshold and no upper limit. Most pronounced effects are in relation to infant mortality, morbidity/mortality from chronic illness (particularly musculoskeletal, CVD, and respiratory conditions), and teenage pregnancy. This is not a new problem nor one unique to the UK. Disparity in health is closely related to income. In the UK, a greater proportion of the population is now living on <50% of average income than 20y ago—the mortality gap has grown proportionately.

Benefits for people with low income ➔ p. 104

Impact of social factors on general practice Factors such as population age, number of people in residential care, high list turnover, rurality, and deprivation have a significant effect on GP workload. This is recognized in the UK by the Carr-Hill Index which adjusts allocation of funds to practices according to these factors (➔ p. 33).

Homelessness

Temporary accommodation Adverse effects of living in temporary accommodation are well documented:

- Adults have an ↑ incidence of depression than people of similar social standing in their own homes
- Homeless women are 2× as likely to have problems in pregnancy and 3× as likely to require admission in pregnancy
- ¼ of babies born to women living in bed and breakfast accommodation are of low birthweight (national average <1 in 10)
- Children from homeless families are less likely to receive their immunizations, more likely to have childhood accidents and have higher incidence of minor respiratory tract and diarrhoeal diseases

Sleeping rough Poor diet, poor accommodation, and lack of access to medical services are universal problems in this group; <70% are registered with a GP. Many homeless people will suffer from a triad of poor physical health, mental health, and substance misuse; homeless people have a higher than average use of emergency and hospital inpatient services as a result.

The average age of death is 40–44y. If primary care services are provided, homeless people do use them.

Divorce Divorcees of all ages are at greater risk of premature death than married people (2× ↑ for men aged 35–42y)—mainly from cardio- and cerebrovascular disease, cancer, suicide, and accidental death. There is also a similar ↑ in morbidity. Children of divorced parents have ↑ risk of ill health from the time of separation until adult life—with children <5y old when their parents separate being particularly vulnerable. These children are also more prone to psychiatric illness later in life and are more likely to become divorced themselves.

Employment and unemployment

> '*Without work all life goes rotten*' Albert Camus

Effects of work have been compared to effects of vitamins—we need a certain amount to be healthy, then there is a plateau where extra does not help—and too much is harmful. There is good evidence that unemployment causes both ↑ mortality (from CVD, cancers, suicide, violence, and accidents) and ↑ morbidity (depression, CVD). Threat of unemployment alone can cause morbidity—in one study, GP consultation rates rose by 20% and referral rates by 60% after it was announced a factory would close. Increases were found in other family members too.

Refugees and asylum seekers The Geneva Convention defines a refugee as any person who, '*owing to well-founded fear of being persecuted for reasons of race, religion, nationality, membership of a particular social group or political opinion, is outside the country of his nationality and is unable or, owing to such fear, is unwilling to return to it*'. Refugees are entitled to free healthcare in the UK. Consider:

- **Language, cultural, and religious issues** (➔ p. 84)
- **Physical health needs** Diverse depending on country of origin and previous level of healthcare. Always consider infectious diseases, e.g. hepatitis B, HIV, TB, and malaria. Ensure refugees claim all health-related benefits available to them (e.g. free prescriptions)
- **Psychological needs** Depression, anxiety, panic attacks, agoraphobia, and poor sleep are common. Symptoms are often reactions to past experiences (including torture) and current situation. Social isolation, unemployment, deprivation, hostility, and racism compound them. Use medication if appropriate but also address other issues. ⚠ Although telling their story is helpful for some refugees, 'active forgetting' is the way some cope
- **Family** Many will have left family members behind. They may not know their whereabouts or even if they are alive. The Red Cross or Red Crescent can help with tracing (🖱 www.redcross.org.uk)

Useful contacts

Freedom from Torture 🖱 www.freedomfromtorture.org
Refugee Council 🖱 www.refugeecouncil.org.uk
Relate Relationship support. 🖱 www.relate.org.uk
Relationships Scotland 🖱 www.relationships-scotland.org.uk
Shelter Help for homeless people. ☎ 0808 800 4444 🖱 www.shelter.org.uk

Multicultural medicine

Britain is a multicultural and multifaith society. It is important that healthcare providers take into account the cultural and spiritual needs of their patients.

❗ Table 4.1 is a rough guide to religious differences that affect healthcare. It is forcibly brief and cannot address all the many variations. Everyone is an individual and there is a real danger of 'pigeon-holing' patients by religion or ethnic background and making incorrect assumptions as a result. Always ask patients/family about their own preferences.

Communication Effective communication is essential. Do not assume English proficiency; it is important to ascertain that you understand the patient and that the patient understands you.

- Ask the patient to let you know if he/she does not understand; consider using an interpreter
- Speak clearly and slowly and repeat important information; avoid jargon, confusing phrases, double negatives, and rhetorical questions
- Ask patients to tell you what you have said to check comprehension
- Be wary of sounding condescending—English skills are not a reflection of a hearing disorder or level of intelligence

Respect beliefs and attitudes People have different reactions towards illness, life, and death. Ask patients to provide you with information about their own ideas, e.g. for newly arrived immigrants, ask: 'Could you tell me what would happen to you if you were in your country?'

Using interpreters Interpreters are an important resource in providing a voice for patients whose proficiency in English is poor or insufficient for the situation. In general, anyone who has been in an English-speaking country for <2y will need an interpreter. Sometimes a friend or another family member can be used but if sensitive issues have to be discussed or it is essential that the information is translated accurately, use a professional. *General tips*:

- Anticipate an interpreter will be needed where possible, and pre-book someone of the same gender who speaks the same language/dialect and will be ethnically acceptable to the patient
- Explain that the interpreter is bound to maintain confidentiality
- Face and speak in the first person directly to the patient, not the interpreter; interpreters are solely there to convey information in a language both patient and doctor can understand—not to analyse information or decide what should or should not be conveyed

Useful contacts

Commercial interpreter services—local PCOs often have contracts with interpreter services for GPs to use during consultations.

Ethnologue Information on languages of the world. 🖱 www.ethnologue.com

MedLine Plus US Government website providing free health information for patients in a wide variety of languages. 🖱 www.medlineplus.gov/languages

Table 4.1 Religious differences important in healthcare

Religion	Dietary restrictions	Fasting	Transfusion/transplant	Family planning	Death
Buddhist	Mainly vegetarian	N/A	No objections	No objections—abortion not allowed	Cremation preferred—no objections to postmortem
Christian	None	N/A	No objections	Some approve of natural methods only	Burial or cremation—no objections to postmortem
Hindu	Most do not eat beef. Some are strict vegetarian	Fasting involves limiting type of foods	No objections	No objections	Strong preference to die at home. The body should not be touched by non-Hindus. All adults are cremated—no postmortems unless legally required
Muslim	No pork. Other meat must have been killed in a special manner (halal). Alcohol is prohibited	Fasting sunrise → sunset during Ramadan	Variable—some Muslims may not consent to transplant	Variable—some Muslims do not approve	All Muslims are buried. No postmortems unless legally required
Jehovah's Witness	No foods containing blood or blood products. No alcohol	N/A	No blood transfusion or organ transplant. Dialysis is usually permitted	No objections	Burial or cremation—no objections to postmortem
Jewish	No pork, rabbit, or shellfish. Meat prepared in kosher fashion. Liberal Jews may not adhere to dietary restrictions	Orthodox Jews may fast for Yom Kippur	No objections	Some orthodox Jews prohibit contraception. Most Jewish boys are circumcised 8d after birth	Burial preferred. No postmortems unless legally required
Sikh	No meat killed in ritualistic fashion. Most are vegetarian. Alcohol is forbidden	N/A	No objections	Allowed but not openly discussed	Children and adults are cremated

Domestic violence: the GP's role

Domestic violence (DV) Any incident or pattern of incidents of controlling, coercive, or threatening behaviour, violence, or abuse between those aged ≥16y who are/have been intimate partners or family members regardless of gender or sexuality. This encompasses, but is not limited to:

- Psychological abuse
- Physical abuse
- Sexual abuse
- Emotional abuse
- Financial abuse

Controlling behaviour Acts designed to make people subordinate or dependent by isolating them from sources of support, exploiting their resources/capacities for personal gain, depriving them of means needed for independence, resistance ± escape, and regulating their everyday behaviour.

Coercive behaviour Act/pattern of acts of assault, threats, humiliation, and intimidation or other abuse used to harm, punish, or frighten victims.

Prevalence Although men may be the victims of DV, ~80% of reported DV is against women by male partners. DV affects ~1 in 4 women and is the most common form of interpersonal crime: 60%—current partner; 21%—former partner. Half suffer >1 attack; 1 in 3 have been attacked repeatedly.

Effects High incidence of psychiatric disorders, particularly depression, and self-damaging behaviours, e.g. drug/alcohol abuse, suicide/parasuicide.

Factors preventing the victim leaving the abusive situation

- Loss of self-esteem makes victims think they are to blame
- Disruption of the family and children's relationship with partner or other key family members (e.g. grandparents, uncles, aunts, cousins)
- Loss of intimate relationship with partner
- Fear of partner
- ↓ in income
- Risk of homelessness
- Fear of the unknown

Presentation General practice is often the first place that victims seek help, but only 1 in 4 actually reveals the true nature of the problem. Without appropriate intervention, violence continues and may ↑ in frequency and severity. By the time injuries are visible, violence may be a long-established pattern. On average, victims are assaulted 35 times before reporting DV to police.

Guidelines for care ❗ Emphasize confidentiality.

- Consider the possibility of domestic violence—ask directly
- Document the patient's story and any injuries—accurate, clear documentation, over time at successive consultations may provide cumulative evidence of abuse and is essential for use as evidence in court, should the need arise
- Assess the present situation—gather as much information as possible
- Provide information; offer help to make contact with other agencies
- Devise a safety plan, e.g. give the phone number of local women's refuge; advise to keep some money and important financial and legal documents hidden in a safe place in case of emergency; help plan an escape route in case of emergency

❗ Do not pressurize the victim into any course of action. If the patient decides to return to the violent situation, in time your information and support might provide the confidence needed to break out of the situation.

⚠ If children are likely to be at risk, inform social services or the police—preferably with the patient's consent.

Elder abuse Single or repeated act, or lack of appropriate action, occurring within any relationship where there is an expectation of trust, which causes harm or distress to an older person. Prevalence is ~4% (↑ with age; ♀:♂ ≈2:1). Older people may report abuse but often do not. Different forms of abuse can be taking place simultaneously (Table 4.2).

Management Talk through the situation with the patient, carer, and other services involved in care. Assess the level of risk. Consider admission to a place of safety—contact social services and/or police as necessary; seek advice from Action on Elder Abuse.

Deprivation of Liberty Safeguards (DoLS) ➔ p. 1106

Assault ➔ p. 88 **Adult safeguarding** ➔ p. 197

Non-accidental injury in children ➔ p. 902

Table 4.2 Elder abuse: what to look for

Type of abuse	Symptoms and signs to look for
General	Patient states he/she has been abused; inconsistent story from patient and carer; inconsistencies on examination; fear shown by the older person in the presence of a carer; frequent attendance at A&E; frequent requests for GP visits; carer avoiding the GP
Physical abuse	Cuts, bruises, unexplained fractures, burns
Psychological abuse	Unusual behaviour, unexplained fear, appears helpless or withdrawn
Financial abuse	Unexplained/unjustified removal of funds by family members, carers, or others; new will in favour of an unexpected recipient
Sexual abuse	Vaginal or anal bleeding, genital infections
Neglect	Malnutrition, dehydration, squalor, poor personal hygiene, late requests for medical attention
Institutional abuse	In a residential care environment, care of the individual is compromised by the rules and routines of the organization, e.g. no food if not hungry when lunch is served

Further information

DH Domestic abuse: a resource for health professionals. 🖱 https://www.gov.uk/government/publications/domestic-abuse-a-resource-for-health-professionals

Useful contacts

Action on Elder Abuse ☎ 0808 808 8141 🖱 www.elderabuse.org.uk
Men's Advice Line ☎ 0808 801 0327 🖱 www.mensadviceline.org.uk
Police, and local authority social services/housing departments
Womens' Aid and National DV Helpline ☎ 0808 2000 247 🖱 www.womensaid.org.uk

Victims of crime

Victims of any crime need treatment of injuries and emotional support.

- Note the date, time, and place of the event
- Record injuries in detail (physical and psychological)—including measuring the size of lacerations and bruises. Record all information carefully as it may be needed for legal cases
- Arrange for photographs to be taken, if appropriate
- Encourage reporting of the incident to the police—the patient will not be eligible for criminal injury compensation if the incident is not reported
- Give patient details of local victim support groups
- If the patient's safety is an issue, contact the duty social worker for a place of safety to be provided

Rape and indecent assault If a patient reports rape or indecent assault and is willing to report the matter to the police, do not perform an examination. The case against the assailant could be won or lost on the basis of evidence gained by examination of an alleged victim, so it is best done by a doctor trained and experienced in such work.

If the patient will not report the matter to the police

- Take a full history of the event. Note: LMP, contraception, sexual history
- Suggest the patient attends a Sexual Assault Referral Centre (SARC) for forensic/medical examination and specialist advice and support

If there is no SARC or the patient is unwilling to attend

- Make a note of any injuries and take photographs if possible and appropriate. Do not insist on examination if the patient is unwilling. Ensure a chaperone is present if any examination is attempted
- Discuss the need for emergency contraception, prophylactic antibiotics (e.g. azithromycin 1g po stat), blood tests at 3mo to exclude transmission of syphilis and at 3–6mo for exclusion of seroconversion for HIV
- If at high risk for HIV transmission, refer to A&E for consideration of prophylaxis (➔ p. 720)
- Discuss the need for counselling, and inform the patient about the victim support scheme and SARCs
- Arrange follow-up in 2–3wk

Domestic violence ➔ p. 86 **Elder abuse** ➔ p. 87

Non-accidental injury in children ➔ p. 902

Modern slavery The Home Office estimates that there are 13,000 victims and survivors of modern slavery in the UK; 55% are ♀ and 35% of all victims are trafficked for sexual exploitation. People are in slavery if they are:

- Forced to work—through coercion, or mental or physical threat. Work may include anything from hard physical work to commercial sexual exploitation or drug trafficking
- Owned or controlled by an 'employer', through mental or physical abuse or the threat of abuse
- Dehumanized, treated as a commodity, or bought and sold as 'property'
- Physically constrained or have restrictions placed on their freedom of movement, e.g. by removing passports

Common types of modern slavery in the UK

- **Debt bondage** Most widespread form of slavery in the UK. People borrow money they cannot repay to come to the UK, and are then required to work to pay off the debt, losing control over the conditions of both their employment and the debt
- **Human trafficking** Involves transporting, recruiting, or harbouring people for the purpose of exploitation, using violence or coercion
- **Forced marriage** When someone is married against their will and cannot leave the marriage

The role of the GP People trapped in modern slavery may come into contact with medical services, particularly in primary care. It is important to be alert to this possibility. In these situations, patients are often not in a position to discuss what is happening to them. As modern slavery is a serious crime, GPs have a duty to report the matter, even if this involves breaking patient confidentiality. Gather as much evidence as you can from your encounter(s) with the patient and consider discussing the issue with your local safeguarding lead and/or the police. There is a national reporting helpline (☎ 0800 0121 700) or modern slavery can be reported online (🖱 https://www.modernslaveryhelpline.org/report).

Prevent Part of the UK's Counter Terrorism Strategy known as CONTEST. Aims to stop individuals from getting involved/supporting terrorism or extremist activity. All healthcare staff must undergo training to recognize signs of radicalization and refer to the police if suspected.

Criminal injuries compensation For victims of violent crimes—even if the attacker is not identified. Compensation is paid for the injury, loss of earnings, and expenses. Claim online or by telephone. ☎ 0300 003 3601 (option 8) 🖱 www.gov.uk/claim-compensation-criminal-injury

Post-traumatic stress disorder (PTSD) 23% of assault victims and 80% of rape victims develop PTSD. ♂:♀ ≈2:1. Defined as significant symptoms 1mo after the event—i.e. flashbacks, nightmares, survivor guilt, mood changes, detachment, poor concentration, insomnia, anxiety, and depression. Alcohol abuse, work, and relationship problems are common. Symptoms may last years. See ➔ p. 976.

Further information

Modern slavery 🖱 www.gov.uk/government/collections/modern-slavery
Prevent Training and Competencies Framework
🖱 www.england.nhs.uk/wp-content/uploads/2017/10/prevent-training-competencies-framework-v3.pdf

Patient information and support

Rape Crisis England and Wales Provides support, information and a list of SARCs and local Rape Crisis Centres. ☎ 0808 802 9999 🖱 www.rapecrisis.org.uk
Rape Crisis Scotland ☎ 0808 801 0302 🖱 www.rapecrisisscotland.org.uk
Survivors UK Provides resources for men who have experienced any form of sexual violence. 🖱 www.survivorsuk.org
Victim Support ☎ 0808 1689 111 🖱 www.victimsupport.org
Victim Support Scotland ☎ 0345 603 9213 🖱 www.victimsupportsco.org.uk

Occupational illness

If a patient develops an occupational disease, a doctor is obliged to notify the employer in writing, with the patient's consent. The doctor does not need to make a judgement about whether the disease is, in that particular case, caused by the occupation.

Employers must then inform the Reporting of Injuries, Diseases and Dangerous Occurrences Regulations (RIDDOR) incident contact centre (☎ 0345 300 9923 🖱 www.hse.gov.uk/riddor). Self-employed patients must contact RIDDOR themselves.

Patients who do not give consent for the doctor to notify their employer may allow the doctor to inform the employer's occupational health department or RIDDOR directly instead.

Industrial injury Injured employees should always report details of any accident to their employer and record them in the accident book as soon as possible—however trivial the injury. Employers must inform RIDDOR of:

- Dangerous incidents—even if no one was hurt
- Incidents where death or serious injury occurs
- Incidents resulting in injury requiring >3d absence from work
- Incidents involving gas

Prescribed industrial disease Disease for which benefit is paid if the applicant worked in a job for which that disease is 'prescribed' and it is likely the employment caused the disease. Claims may be made at any time with the exceptions of occupational deafness (claim <5y after leaving employment) and occupational asthma (claim <10y after leaving employment).

Notifiable industrial diseases ❗ This is not a complete list:

- Poisoning by industrial agents, e.g. lead, arsenic, mercury
- Bursitis, e.g. housemaid's knee
- Occupational asthma
- Folliculitis and acne (associated with work with tar, pitch, or oils)
- Occupational infection, e.g. hepatitis B in healthcare workers, anthrax in farmers
- Chrome ulceration
- Irritant dermatitis, e.g. hair-dressers' dermatitis
- Vibration white finger
- Repetitive strain injury
- Tenosynovitis, e.g. as a result of repeated movements of the hand/wrist
- Pneumoconiosis
- Extrinsic allergic alveolitis
- Occupational deafness
- Occupational cancers, e.g. nasopharyngeal cancer in woodworkers, bladder cancer in plastic workers, cancers as a result of ionizing radiation, mesothelioma due to asbestos exposure

The list of prescribed diseases is similar to, but not the same as, the list of notifiable diseases.

Making claims Through local Industrial Injuries Disablement Benefits offices. A full list of prescribed industrial diseases is also available from these offices. Some claims can be made online. For further information contact ☎ 0800 121 8379 🖱 www.gov.uk.

Industrial benefits that may be payable

Industrial Injuries Disablement Benefit Available to employed earners for injuries resulting from accidents or certain (prescribed) illness arising as a result of employment, even if the employee was either part or wholly to blame. 'Industrial' covers virtually all forms of work. For accidents, claims can be made at any time after the event but benefit is paid only if there are still effects of the injury after the 91st day.

Payable if the person was a paid employee at the time of the accident or when he/she contracted the disease; *and* disability is assessed at ≥14% (exceptions: occupational deafness >20%; dust-related lung disease—no level). If a patient claims benefit for >1 industrial accident or disease, assessments may be added together and benefit awarded on the total.

Reduced Earnings Allowance Payable if the accident occurred or disease was contracted prior to 1 October 1990, the disability is assessed at ≥1%, and the individual is

- Unable to work, *or*
- Unable to do his/her normal job or another job with equivalent pay, *or*
- Working less hours at his/her normal job

Retirement Allowance Reduced earnings allowance becomes retirement allowance at statutory age of retirement. It is paid at 25% the rate of reduced earnings allowance when a claimant stopped work.

Constant Attendance Allowance For people who need daily care and attention and who are getting industrial injuries disablement benefit for disability assessed at 100% or 80% war disablement pension. Four rates of benefit depending on the level of care required.

Exceptionally Severe Disablement Allowance For people who get constant attendance allowance at 'exceptional' or 'intermediate' rate and where need for attendance is likely to be permanent.

❗ People who suffer from industrial diseases or have suffered disability as a result of an industrial accident are also eligible to apply for benefits available for any disabled individual (➔ p. 108).

Benefits for service veterans ➔ p. 106

Further information

Citizens Advice 🖱 www.adviceguide.org.uk
RIDDOR Incident Contact Centre. ☎ 0345 300 9923 🖱 www.hse.gov.uk/riddor
Trade Unions

Time off work

In the UK, 31.2 million working days were lost due to work-related ill health and workplace injuries in 2016/17. Average time off work was 17d. The most common reasons were:

- Stress, depression, and/or anxiety (12.5 million days lost)
- Musculoskeletal problems (8.9 million days lost)

Facts and figures

- Sickness absence ↑ with age; ♀ have higher rates than ♂
- There has been a ↓ in amount of sick leave over the past 20y
- 16% of sick leave is for >20d, but this accounts for 32% of lost time
- The longer someone is not working, the less likely that person is to return to work; someone who has been off sick for ≥6mo has an 80% chance of being off work for 5y

Benefits of returning to work Going back to work promotes recovery, ↑ physical/mental health and well-being, and ↓ social exclusion/poverty. In contrast, long periods out of work can cause/contribute to:

- ↑ consultation, medication consumption, and hospital admission rates
- 2–3× ↑ risk of poor general health and mental health problems
- 20% excess mortality

The role of the GP When someone of working age presents with a problem that affects ability to work, record a brief occupational history:

- Address the underlying health problem and any personal, psychological, organizational, or social factors preventing return to work
- Wherever possible, suggest work adjustments where appropriate to enable a patient to return to work (e.g. graduated work or transitional arrangements) or instead of signing the patient off work; do this through the 'remarks' section of the 'Statement of Fitness to Work'—➔ p. 95
- Involve occupational health professionals if possible

Certification of time off work ➔ p. 95

Postoperative time off work Table 4.3

Time off work for emergencies In many cases, patients have the legal right to take time off work to deal with an emergency involving someone who depends on them. They may only be absent for as long as it takes to deal with the immediate emergency; employers do not have to pay for their time.

Dependants Include spouse or partner, children, parents, or anyone living with the patient as part of their family. Others who rely wholly on the patient for help in an emergency may also qualify.

Emergencies Include situations in which a dependant:

- Is ill and needs help
- Goes into labour
- Is involved in an accident or assaulted
- Needs the patient to arrange their longer-term care
- Needs the patient to deal with an unexpected disruption or breakdown in care, such as a childminder or nurse failing to turn up
- Dies and the patient must make arrangements/attend the funeral

Table 4.3 Expected postoperative time off work for common surgical procedures

Surgical procedure	Time off work (wk)	
	Minimum	Maximum for uncomplicated procedures
Angiography/angioplasty	<1	4
Appendectomy	1	3
Arthroscopy (knee)	1	4
Cataract surgery	<1	2
CABG or valve surgery	6	12
Cholecystectomy	**2** (laparoscopic)	**12** (open)
Colposcopy ± cautery	<1	<1
Cystoscopy	<1	<1
ERPC or surgical TOP	<1	<1
Femoro-popliteal grafts	4	12
Haemorrhoid banding	<1	<1
Haemorrhoidectomy	2	4
Hysterectomy	**2** (laparoscopic)	**8** (open)
Inguinal or femoral hernia repair	**1** (laparoscopic)	**6** (open)
Laparoscopy ± sterilization	<1	<1
Laparotomy (open)	6	12
Mastectomy	2	12
Pacemaker insertion[a]	<1	<1
Pilonidal sinus[b]	2	8
Retinal detachment	<1	Avoid heavy work life-long
Total hip/knee replacement	6	26
TURP	2	8
Vasectomy	<1	2

[a] Driving rules following pacemaker insertion ➔ p. 243

[b] If time off work is allowed for dressings

❗ These are not hard and fast rules—alter them to fit individual circumstances (e.g. laparoscopic procedures often entail less time off than open procedures; patients performing hard manual jobs may require more time off work).

Certifying fitness to work

Individuals must self-certify for the first 7d of incapacity, then sickness certification from a GP is needed until the Work Capability Assessment (WCA) is carried out.

Own occupation test Applies to those claiming Statutory Sick Pay (SSP) for the first 28wk of illness. The GP assesses if the patient is fit to do his/her *own* job.

Work Capability Assessment Carried out by employment advisers contracted to the Department for Work and Pensions (DWP). It is not diagnosis dependent and assesses a variety of different mental/physical health dimensions for ability to work. WCA is performed within the first 13wk of any claim for Employment Support Allowance (ESA) or Universal Credit and applies to:

- Everyone after 28wk incapacity
- Those who do not qualify for the 'own occupation test' from the start of their incapacity (i.e. do not qualify for SSP)

Initial information All applicants are asked to fill in the 'Limited capability for work' questionnaire which explores how the individual's medical problem impacts ability to work. Sometimes medical reports from GPs may be sought by the DWP at this stage. GPs have a contractual obligation to complete and return these reports.

Medical examination In the majority of cases, more information will be needed to be able to assess the claim, and the claimant is then invited for a face-to-face medical examination assessing mental and physical ability to work. Groups considered unfit to work without medical examination include pregnant women, people with severe physical or learning disability, and those who are terminally ill.

Classification Based on these assessments people may be placed into one of 3 groups: fit to work, work-related activity group, or support group.

Those placed in the *work-related activity group* take part in work-focused interviews with personal advisers and are provided with a range of support to help them prepare for a return to work.

Those placed in the *support group* have an illness/disability that has a severe effect on ability to work. They are not expected to take part in any work-related activity but can choose to do so if they wish.

Appeals Once a decision has been made about ESA/Universal Credit, the individual's GP is informed and no further sickness certification is needed. If the claimant disagrees with the decision, he/she can ask for it to be reconsidered, and if that fails to alter the decision, can appeal. GPs must continue sickness certification pending the appeal decision.

Equality Act 2010 An employer has to make 'reasonable adjustments' to avoid an employee with an ongoing health problem/disability being put at a disadvantage (e.g. adjusting working hours or providing equipment).

Forms for certifying incapacity to work

SC1 Self-certification form for people not eligible to claim SSP who wish to claim ESA/Universal Credit. Certifies first 7d of illness. Available from local benefits offices and GP surgeries.

SC2 As SC1 but for people who can claim SSP. Available from employers, local benefits offices, and GP surgeries.

Statement of Fitness for Work (Med3) Filled in by a GP or hospital doctor who knows the patient for periods of incapacity likely to be >7d. In general practice, usually computer generated and saved directly to the patient's medical record.

During the first 6mo of incapacity can only be issued for a maximum period of 3mo. Gives the doctor two options:

- The patient is unfit for work
- The patient may be fit for work—this allows the GP to recommend circumstances under which the patient may be able to return to work, e.g. with restricted duties, workplace adaptions, or reduced hours

The form gives space for the GP to record the patient's functional limitations. This is designed to allow the employer to make adjustments to facilitate the employee's return to work. The GP can also mark on the form whether there is an intention to review the patient again before return to work.

The Statement of Fitness for Work may be issued:

- On the day of your assessment of the patient (telephone consultations are acceptable)
- On a date after your assessment of the patient if you think that it would have been reasonable to issue a Statement on the day of your assessment
- After consideration of a report or other medical record about the patient from another doctor or registered healthcare professional

Only one Statement of Fitness for Work can be issued per patient per period of sickness. If mislaid, reissue and mark 'duplicate'.

❗ Employers may not request certification that employees 'need not refrain from work'—if required, this should be requested as a private service from an occupational health physician or GP.

Mat B1 Signed by doctor or midwife. Provided to pregnant women once within 20wk of estimated date of delivery. Enables her to claim statutory maternity pay and other benefits (➜ p. 765).

Private certificates Some employers request a private certificate in the first week of sickness absence. They should request it in writing. If the GP chooses to provide the service, a charge can be made both for a private consultation and the provision of a private certificate.

Further information

DWP (2016) Fit note: guidance for GPs. 🖱 www.gov.uk/government/publications/fit-note-guidance-for-gps
Gov.uk Disability rights: employment. 🖱 www.gov.uk/rights-disabled-person/employment

Fitness to make decisions

Mental capacity The ability to take actions affecting daily life (e.g. when to get up, what to wear, what to eat) and/or make more major decisions (e.g. where to live, how to manage money).

Mental Capacity Act (2005) Came into force in 2007 in England and Wales. Similar legislation applies elsewhere in the UK. It specifies who can take decisions on behalf of other people and allows people to plan ahead for a time when they may lack capacity. 5 key principles:

1. Every adult has the right to make decisions and must be assumed to have capacity to make them unless proved otherwise
2. Every adult must be given all possible help and support to make decisions, and to communicate those decisions where necessary, before he/she can be assumed to have lost capacity
3. Making an unwise decision does not mean that a person lacks capacity to make that decision
4. Anything done or any decision made on behalf of someone who lacks capacity must be done in his/her best interests
5. Anything done or any decision made on behalf of someone who lacks capacity should be the least restrictive of his/her basic rights/freedoms

Assessing capacity

- Have access to the patient's records and ideally know the patient
- Seek information from friends, relatives, carers, and/or the patient's independent mental capacity advocate, if one has been appointed
- Examine the patient and assess the type and degree of deficit
- Decide if there is an impairment of, or disturbance in, the functioning of the patient's brain or mind
- If there is a disturbance, decide if the patient is able to make the particular decision in question—in particular:
    - Can the patient understand the relevant information, including the likely consequences of making/not making that decision?
    - Can the patient retain that information?
    - Can the patient use or weigh that information as part of the process of making the decision?
    - Can the patient communicate that decision by any means?
- Decide if assessment should be postponed while measures are taken to improve capacity
- Record all the above-listed information

❗ Even if a proposed action is in the patient's best interests, do not judge the patient capable if not clearly the case. Seek a second opinion if in doubt.

Lasting Power of Attorney (LPA) Replaced Enduring Power of Attorney (EPA) in October 2007. However, people with EPAs in place can still use them. An LPA is a legal document that lets individuals appoint someone they trust to make decisions for them. It can be drawn up at any time whilst the person has capacity but has no legal standing until it is registered with the Office of the Public Guardian. Two types:

Property and affairs LPA Allows the 'attorney' to make decisions about the management of money, property, and affairs. Unless specified otherwise, can be used even when the individual retains capacity.

Personal welfare LPA Allows the 'attorney' to make decisions about healthcare and welfare, including decisions to refuse or consent to treatment, and decide on place of residence. Only active when the LPA is registered and the individual lacks capacity to make decisions. The attorney can make decisions about life-sustaining treatment only if the LPA specifies that.

Court of Protection If a person, by reason of mental disorder, becomes incapable of managing his or her affairs but has not previously signed an LPA, it may be necessary for someone, usually the nearest relative, to apply to the Court of Protection for the appointment of a 'receiver' to do so. The medical practitioner will be asked to complete form CP3. Alternatively, if the patient's affairs are simple (e.g. state pension), direct arrangements can be made with the relevant authorities.

Testamentary capacity The capacity to make a will. Anyone can make a will provided they understand the nature and effect of making a will, extent of property being disposed of and claims others may have on that property, and the decision is not the result of their condition (e.g. due to a delusion).

❗ Decisions do not have to seem rational to others, especially if consistent with pre-morbid personality.

Consenting to medical treatment ➔ p. 48

Advance decisions Statements about wishes regarding medical treatment in case the individual becomes incapable of making that decision later. Advance decisions are legally binding.

- Respect any refusal of treatment as long as the decision is clearly applicable to circumstances, there is no reason to believe the individual has altered that decision, and the decision was not made under duress
- Advance decisions do not have to be written, except those refusing life-sustaining treatment which must be: specific to a particular treatment (e.g. refusal to have CPR); written; signed by the person making the decision (or a representative if unable to sign) and a witness
- Advance decisions cannot include decisions about treatment the person would like, only treatment the person refuses, and cannot include directions to end the person's life prematurely
- Doctors may not be willing to carry through an advance directive. In such cases they should refer the patient to another doctor who is
- The BMA recommends doctors should not withhold 'basic care' (e.g. symptom control), even in the face of a directive which specifies that the patient should receive no treatment
- Where a formal advance statement is not available, take patients' known wishes into consideration

Deprivation of Liberty Safeguards (DoLS) ➔ p. 1106

Further information

Office of the Public Guardian 🖱 www.publicguardian.gov.uk
Medical defence organizations

Fitness to drive

⚠ Driving licence holders (and applicants) have a legal duty to inform the DVLA of any disability likely to cause danger to the public if they drove.

Driving licence types

Group 1 Ordinary licence for driving a car/motorcycle. Minimum age 17y (16y if disabled). Old licences expire at 70th birthday and then must be renewed 3-yearly. Applicants are asked to confirm they have no medical disability. If so, no medical examination is necessary. New photocard licences are automatically renewed 10-yearly until age 70y.

Group 2 Licences enable holders to drive lorries and buses. Minimum age 21y. Initially valid until 45th birthday then renewable by medical examination every 5y until 65th birthday. >65y renewable annually. Applicants must bring form D4 (available from post offices) with them. Examinations take ~½h. A fee may be charged by the GP.

Determining fitness to drive Patients with any disorder which may cause danger to others if they drove should be advised not to drive and to contact the DVLA. The DVLA gives advice on when they can restart.

Breaching confidentiality When a patient continues to drive despite advice by a doctor to stop, a doctor has an obligation to breach confidentiality and inform the DVLA.

- If the patient does not understand the advice to stop driving—inform the DVLA immediately
- If the patient does understand, but continues to drive—explain your legal duty to inform the DVLA. If the patient still continues driving, offer to refer to a colleague for a second medical opinion—on the understanding that the patient stops driving in the interim. If all else fails, inform the DVLA in confidence. Before doing this, inform the patient of your intended actions and consider contacting your medical defence organization for advice. Once the DVLA has been informed, you should also write to the patient to confirm disclosure has been made

Visual acuity Drivers must be able to read in good light (with glasses or contact lenses) a number plate containing figures 79mm high × 57mm wide at a distance of 20.5m (20m where the characters are 50 mm wide). In addition, Group 2 drivers must have corrected vision of ≥6/9 (best eye) and ≥6/12 (other eye); they should not drive if uncorrected acuity in either eye is <3/60.

Condition-specific guidance For UK drivers, the DVLA provides detailed condition-specific guidance about fitness to drive. Information about driving is included with the relevant clinical topics in this book. However, as the DVLA standards are updated every 6mo, always check the most recent version of the DVLA *Assessing fitness to drive: a guide for medical professionals* before giving patients advice.

Multiple medical conditions A combination of medical conditions, each insufficient itself to disqualify from driving, may together render a person unfit or unsafe to drive. If this is the case, advise the patient not to drive and seek clarification from the DVLA.

⚠ **Medication** It is an offence to drive or attempt to drive while unfit through the effect of drugs; the law does not distinguish between illicit and prescribed drugs. GPs prescribing and/or dispensing medication that affects ability to drive should advise patients of that risk.

Driving after surgery Drivers do not need to notify the DVLA following surgery unless a condition likely to affect safe driving persists >3mo (certain exceptions apply for neurological and cardiovascular disorders). It is the responsibility of the driver to ensure that he/she is in control of the vehicle at all times. It might also be advisable for the driver to check with his/her insurer before returning to driving after surgery. Consider:

- Recovery from anaesthesia (sedation and cognitive impairment)
- Impairment due to analgesia (sedation and cognitive impairment)
- Physical restrictions due to the surgery or the underlying condition

Disabled drivers Disabled people who want to learn to drive, or return to driving following onset of their disability, should have an assessment of their driving ability and/or advice on controls and adaptations needed. Licences may be limited to adapted vehicles. A list of driving assessment centres can be obtained from Driving Mobility (☎ 0800 559 3636 🖱 www.drivingmobility.org.uk).

Seat belt exemption GPs can sign a form to exempt patients (e.g. those with colostomies) from having to wear a seat belt. Consider very carefully the reasons for exemption in view of the weight of evidence in favour of seat belts preventing serious harm or injury in the event of collision.

Further information

Department for Transport Medical exemption from compulsory seat belt wearing: guidance for medical practitioners. 🖱 www.gov.uk/government/publications/medical-exemptions-from-compulsory-seat-belt-wearing/medical-exemption-from-compulsory-seat-belt-wearing-guidance-for-medical-practitioners

Driving Mobility ☎ 0800 559 3636 🖱 www.drivingmobility.org.uk

DVLA Assessing fitness to drive: a guide for medical professionals. 🖱 www.gov.uk/government/publications/assessing-fitness-to-drive-a-guide-for-medical-professionals

DVLA Medical advisers from the DVLA can advise on difficult issues—contact: The Medical Adviser, Drivers Medical Group, DVLA, Swansea SA99 1DA or ☎ 01792 782337 (in Northern Ireland: ☎ 0300 200 7861) or E-mail: medadviser@dvla.gsi.gov.uk (medical professionals only)

Patient information

Gov.uk Driving and medical conditions. 🖱 https://www.gov.uk/transport/driving-and-medical-conditions

Fitness for other activities

⚠ Remember—signing a form may result in legal action against you should the patient *not* be fit to undertake an activity.

- Where possible, include a caveat, e.g. 'based on information available in the medical notes, the patient appears to be fit to … although it is impossible to guarantee this'
- If unsure, consult your local LMC or medical defence organization for advice

Fitness to fly Passengers are required to tell the airline at the time of booking about any conditions that might compromise their fitness to fly. The airline's medical officer must then decide whether to carry them.

Hazards of flying

- Cabin pressure—oxygen levels are lower than at ground level and gas in the body cavities expands 30% in flight
- Inactivity and dehydration
- Disruption of routine
- Alcohol consumption
- Stress and excitement

Contraindications to flying Table 4.4

Precautions

- Carry all regular medication especially relief medications (e.g. salbutamol, GTN spray) in the cabin
- For people who have to time their medication carefully, keep to the times that medication was taken at home for duration of flight, e.g. for patients with DM—take snacks to eat and take insulin at normal times
- Drink plenty of liquid (non-alcoholic) to prevent dehydration
- Do calf exercises/get up and walk up and down at intervals to prevent venous stasis in the legs—those at risk of venous thromboembolism should wear compression stockings for the flight
- Pre-warn airlines of special needs so that they can accommodate them, e.g. extra leg room, special diet, oxygen in-flight, transport to/from the plane

Further information

Civil Aviation Authority (CAA) Am I fit to fly? 🖱 https://www.caa.co.uk/Passengers/Before-you-fly/Am-I-fit-to-fly-/

Pre-employment certification It is becoming increasingly common for GPs to be asked about the 'medical' suitability of candidates to perform a job. This is not part of the GP's terms of service and therefore a GP can refuse to give an opinion. In all cases where an opinion is given, a fee can be claimed. Common examples are:

- Forms for childminders
- Care home staff—proof of 'physical and mental fitness'
- Food handlers—certificates of fitness

Table 4.4 Contraindications to flying

Type of condition	Contraindications to flying
Respiratory disease	*Suspected pneumothorax/pneumomediastinum*—patients should not fly for 14d after complete resolution of pneumothorax *Chronic lung disease*—if a patient can walk >50m or climb a single flight of stairs without significant breathlessness, he/she should be fit to fly. Supplementary oxygen can be provided in flight for patients unable to walk this far but the patient must pre-book this with the airline and there is usually a fee
Heart disease	Patients should not travel if they have unstable angina, poorly controlled heart failure, or an uncontrolled arrhythmia, Patients should also refrain from travelling <10d after uncomplicated MI (3–4wk if complicated recovery) and for 3–5d after angioplasty
Thromboembolic disease	Patients should not travel with a DVT before established on anticoagulants
Neurological disease	Patients should not travel for 3d after stroke, or, if epileptic, <24h after a grand mal fit
Infectious disease	Patients must not travel with untreated infectious disease
Psychiatric disease	Patients should not travel if they have disturbed or unpredictable behaviour that could disrupt the flight
Fractures	Flying is restricted for 24–48h (depending on the length of the flight) after the plaster cast has been fitted
Haematological disease	Anaemia (<7.5g/dL) and recent sickling crisis may restrict flying
Pregnancy	Most airlines will not carry women >36wk pregnant (3rd trimester if multiple pregnancy) or with history of premature delivery, cervical incompetence, bleeding, or ↑ uterine activity
Babies	<2d old should not fly (preferably <7d old)
Ear problems	Flying with otitis media or sinusitis can result in pain ± perforation of the ear drum. Patients are advised not to fly until symptoms resolve
Surgery	Patients should not travel <10d after surgery to the chest, abdomen, or middle ear. Other procedures where gas is introduced into the body also need careful consideration

Fitness to perform sporting activities ➔ p. 474

Bereavement, grief, and coping with loss

Models of grief In the traditional model, the bereaved person moves through phases until 'recovery':

- **Initial shock** Sense of unreality, detachment, disbelief, or 'numbness'. Lasts from hours to days
- **Yearning** Pangs of grief, episodes of intense pining, and a desire to search interspersed with anxiety, guilt, and self-reproach
- **Despair** The permanence of the loss is realized. Characterized by despair, apathy, social withdrawal, poor concentration, and pessimism about the future
- **Recovery** Rebuilding of an identity and purpose in life

Recent models Newer models of grief are less linear: grief represents an oscillation between loss- and restoration-focused behaviour, demonstrated by swings in mood, thoughts, and behaviour between memories of the dead person and 'getting on with life'. Avoidance or denial of the loss is common and a part of the process.

Health consequences of bereavement

- **↑ mortality** (↑ deaths from CVD, cirrhosis, suicide, accidents) particularly in first 6mo. Risk factors: ♂ > ♀, age <65y, lower social class
- **Mental health problems** Depression, anxiety, ↑ risk of suicide, substance abuse, identification reaction (hyperchondriacal disorder—symptoms mimic those of deceased, e.g. chest pain if died from MI), insomnia, self-neglect
- **Physical problems** Fatigue, aches, and pains (e.g. headaches, musculoskeletal pain), appetite change, GI symptoms, ↓ immune response (↑ minor infection)
- **Others** Interference with family life, education and employment, social isolation/loneliness, ↓ income

Role of the primary care team Develop a practice policy for bereaved patients. Flag notes. Consider staff training and active follow-up of bereaved patients. If the person who has died is registered with the practice, ensure all medical referrals/appointments are cancelled.

Bereaved children Children understand what death is by 8y, and even children of 2–3y have some understanding of death. Exclusion makes children isolated and often makes the death of someone they have known more, not less, painful. Prepare children for a death if possible and give them a chance to have their questions answered. If a child has problems, seek specialist help.

Bereavement Support Payment Payable if husband, wife, or civil partner dies aged < state pension age, and either has paid enough National Insurance contributions, or death was caused by employment. Consists of:

- Initial lump sum, and then
- Up to 18 monthly payments

Two rates. The higher rate is payable if there are children <16y (or <20y in full time education) in the home or a woman whose partner has died is pregnant.

Claims forms can be obtained from JobcentrePlus offices or downloaded from 🖱 www.gov.uk/bereavement-support-payment. To receive the full amount, claims should be made <3mo after the death (although will be accepted for up to 21mo). Further information can be obtained from ☎ 0800 731 0469.

Budgeting Loans for funeral payments ➔ p. 105

Deaths of service personnel Where someone dies during service, Veterans UK automatically considers whether the death was due to service and whether benefits are payable to any dependants. If ex-service personnel die as a result of illness/injury sustained while in service, compensation may also be payable. Except for rare exceptions, the Armed Forces Compensation Scheme generally only applies if someone dies <7y after leaving the services. Claims must be made <3y after the death. Further information can be obtained from: ☎ 0808 1914 2 18 🖱 www.gov.uk/veterans-uk.

Abnormal grief reactions Whether a grief reaction is normal or abnormal depends on individual circumstances—personality, situation surrounding death, and cultural expectations. Recognized patterns of abnormal grief include:

- Inhibited grief—grief is absent or minimal
- Delayed grief—late onset
- Prolonged or chronic grief—inability to rebuild life in any way

If abnormal grief is suspected Monitor carefully. Consider referral for bereavement counselling, e.g. to CRUSE. Consider clinical depression (➔ p. 978) or post-traumatic stress disorder (➔ p. 976). If symptoms are persistent or worsening despite treatment or if there is suicidal risk, refer to the mental health team for specialist advice.

Risk factors for poor outcome after bereavement

Predisposing factors

- Multiple prior bereavements
- Poor social or family support
- Low self-esteem
- Being male
- History of mental illness, e.g. depression, anxiety, self-harm
- Ambivalent or dependent relationship with the deceased

Factors associated with the nature of the death

- Sudden or unexpected death
- Suicide
- Death due to unnatural causes e.g. murder, road accident, drowning
- Multiple deaths e.g. natural disaster, terrorist attack
- Avoidable deaths e.g. drug overdose, missed diagnosis
- Death of a child (or for children, death of a parent or sibling)
- Feelings of guilt e.g. if driver of a car involved

Patient information

CRUSE Bereavement Care ☎ 0808 808 1677 🖱 www.cruse.org.uk
National Association of Widows ☎ 0845 838 2261 🖱 www.nawidows.org.uk

Benefits for people on low income

Benefit cap Limits the total amount of state benefit that most people aged 16–64y can receive. The amount of the cap varies according to where the person lives and personal circumstances.

Income Support (IS) can be claimed by people aged ≥16y and:

- Pregnant, a carer, or a single parent looking after a child <5y, or unable to work due to sickness or disability, or aged 16–20y and in full-time education/training (excluding higher education)
- Working <16h/wk (some voluntary work may be allowable)
- Under Pension Credit qualifying age (➜ p. 106)
- On a low income or have no income
- Have <£16,000 in savings

Jobseeker's Allowance (JSA) Can be claimed by people aged ≥18y and under state pension age who are unemployed or working <16h/wk. Claimants must be: available to work full-time, actively looking for full-time work, not in full-time education and not claiming IS. There are 2 types of JSA:

- Contribution based—for people who have worked, paid sufficient National Insurance (NI) and have not claimed JSA in the past 2y. Payable for 6mo
- Income based—for people who have not paid sufficient NI or have made a JSA claim in the past 2y. Eligible if savings <£16,000, partner (if applicable) is working <24h/wk, and not claiming Pension Credit, Employment Support Allowance, Universal Credit, or IS

Tax Credits Two types:

- Working Tax Credit (WTC)—for people on low income working ≥16h/wk (>30h/wk if aged 25–59y and no childcare responsibilities)
- Child Tax Credit (CTC)—for people with children <16y (or 16–20y and in full-time education/training) who are working and on low income. A credit is payable for each child born before 1.4.2017; if children are born after that date, credits are only payable for 2 children per family. Additional allowances are made for disabled children

Pension Credit ➜ p. 106

Child Benefit ➜ p. 827

Housing Benefit Payable to families on low income to pay rent. Only 1 member of a couple can apply. People with >£16,000 in savings are not eligible. If savings of £6000–£16,000, Housing Benefit may be paid at reduced rate.

Universal Credit Benefit first introduced in October 2013 to replace:

- Income Support
- Child Tax Credit
- Working Tax Credit
- Income-related Employment and Support Allowance
- Income-based JSA
- Housing Benefit

❗ Universal credit is being rolled out across the UK with the aim that all new claimants will receive Universal Credit rather than 'legacy benefits' by 2022.

Local authority payments Council Tax Reduction, Community Care Grants, and Crisis Loans for general living expenses have been replaced with payments from local authorities. Local schemes vary.

Short-term advances Provided by the DWP if financial hardship because of issues with benefit payments.

Budgeting Loans Can help pay for:

- Furniture
- Household items, e.g. washing machine
- Clothes/footwear
- Costs of moving home
- Rent advances
- Home maintenance
- Home security
- Costs of job hunting
- Maternity costs
- Funeral costs
- Repayment of certain loans

Applications are made online. Only people claiming IS, income-based JSA, income-related Employment Support Allowance, and Pension Credit for >6mo can request budgeting loans. People claiming Universal Credit must apply for a Budgeting Allowance instead.

Automatic health benefits People on certain low-income benefits ('passport' benefits) may be able to claim:

- Free NHS prescriptions (➔ p. 113)
- Free NHS dentistry, eye tests/glasses, wigs and fabric supports
- Some hospital travel costs
- Milk, vitamins, and fresh fruit/vegetables for pregnant and breastfeeding women and children <5y
- Free school meals

❗ Passported benefits gradually ↓ as income ↑.

Cold weather payments Paid automatically to people with low income, or on retirement pension, if the temperature is <0°C for ≥7 consecutive days.

Food banks Provide emergency food supplies on presentation of a voucher. Arrangements for provision of vouchers vary across the UK, but generally food vouchers are issued by Jobcentre Plus offices, social workers, Citizen's Advice, and some GP surgeries.

Benefit fraud Occurs when people deliberately claim benefits they are not entitled to, e.g. by providing inaccurate information or not reporting any change in circumstances. Suspected benefit fraud can be reported online via 🖱 www.gov.uk or via the National Benefit Fraud Line ☎ 0800 854 4400.

Further information

Budgeting Loans 🖱 www.gov.uk/budgeting-help-benefits
Child Tax Credit 🖱 www.gov.uk/child-tax-credit
Citizen's Advice 🖱 www.adviceguide.org.uk
Housing Benefit 🖱 www.gov.uk/housing-benefit
Income Support 🖱 www.gov.uk/income-support
Jobcentre Plus ☎ 0800 055 6688 🖱 www.gov.uk
Jobseeker's Allowance 🖱 www.gov.uk/jobseekers-allowance
Universal Credit ☎ 0800 328 5644 🖱 https://www.gov.uk/browse/benefits/universal-credit
Working Tax Credit 🖱 www.gov.uk/working-tax-credit

Pensions

Retirement pension A state retirement pension is currently payable to people of state pension age even if still working. Claim forms should be received automatically—if not, request one through the local Jobcentre Plus office. Pensions are taxable. State pension age is gradually increasing—in women from 60 to 67y and in men from 65 to 67y.

Basic pension Flat rate amount—different for single people and married couples. If not enough NI contributions have been paid, amounts may ↓. >80y, a higher rate is payable which is not dependent on NI contributions.

Variations on basic pension

- Increase for dependants—if spouse/partner is <60y and on low income
- Additional pension—second state pension for ♂ born before 6.4.1951 and ♀ born before 6.4.1953. Based on earnings, NI contributions, and additional pension contributions through the State Earnings-Related Pension Scheme (SERPS) (1978–2002) and/or State Second Pension Scheme (2002–2016)
- Graduated pension—based on earnings from 1961 to 1975
- Extra pension—for people who defer claiming their basic pension for up to 5y. Extra pension is payable when pension is eventually claimed

Pension Credit Tops up state pension for pensioners with weekly income below a threshold amount. Threshold varies according to factors such as whether the person lives alone or is part of a couple, savings, and other benefits received. Receipt of Pension Credit confers automatic eligibility for Housing Benefit (➔ p. 104) and Budgeting Loans (➔ p. 105). Claims can be made via the Pension Credit Claim line (☎ 0800 99 1234).

National insurance credits Protect Basic State Pension for people who do not work because of illness, unemployment, child care responsibilities, or because they are carers.

Christmas bonus One-off payment made to people receiving a retirement pension a few weeks before Christmas.

Other benefits just for pensioners

- Free colour TV licence All pensioners >75y; currently under review
- Winter fuel payment Annual payment to all pensioners >60y

Cold Weather Payment Paid automatically to people with low income or on retirement pension if the temperature is below freezing for 7 consecutive days.

Benefits for:

- Low income ➔ p. 104
- Disability ➔ p. 108
- Bereavement ➔ p. 102

Maintaining independence ➔ p. 198

Pensions for ex-Service personnel

War Pensions Scheme For personnel who have served in the British armed forces and whose injuries and illnesses are related to their service, and arose prior to 6 April 2005. No time limit for claims. Administered by the Veterans Agency. Includes:

- **War Disablement Pension** Based on percentage disability and may be a one-off lump sum (if <20% disabled) or paid as a regular weekly sum (pension). In addition, pensioners may be awarded a mobility supplement for walking difficulty (holders can apply for the Motability Scheme and Road Tax Exemption—➔ p. 199) and/or a Constant Attendance Allowance (➔ p. 91) if high levels of care are needed
- **Medical treatment** Some services and appliances may be paid for by the Veterans Agency (includes prescription charges, nursing home fees)
- **War Widows and Widowers' Pensions** For spouses/civil partners of Service and ex-Service personnel where death was related to service; an additional allowance is paid on reaching the age of 65y with further increases at 70y and 80y

Armed Forces Compensation Scheme (AFCS) Administered by the Veterans Agency. Provides benefits for illness, injury or death caused by service in the armed forces on or after 6 April 2005. Time limit for claims is 7y from the event, from the time when medical advice was first sought or after retirement—whichever is soonest. There is an exceptions list for late-onset conditions. Provides:

- Lump sum for significant illnesses/injuries—15 levels of award
- Tax-free Guaranteed Income Payment (GIP) for life for injuries at the higher tariff levels (1–11) to compensate for loss of earnings capacity
- Guaranteed Income Payment for Survivor's (SGIP) where an attributable death occurs

Patient information

Citizens Advice 🖱 www.adviceguide.org.uk
Pension Service ☎ 0800 99 1234 🖱 www.gov.uk/state-pension
Royal British Legion ☎ 0808 802 8080 🖱 www.britishlegion.org.uk
SSAFA The Armed Forces Charity. ☎ 0800 731 4880 🖱 www.ssafa.org.uk
Veterans Agency ☎ 0808 1914 218 🖱 www.gov.uk/veterans-uk

Benefits for sickness and disability

Benefits for people unable to work due to illness

Statutory Sick Pay (SSP) Weekly sum paid through the employer's normal payroll mechanisms from 3d into illness and for up to 28wk to employees who are:

- Age ≤16y and under state pension age
- Incapable of work due to sickness or disability
- Earning ≤ the NI lower earnings limit (in 2018/19, £116/wk), and
- Unable to work for ≤4d consecutively (including days when the person would not normally work)

Claimants can self-certify that they are unfit to work for the first week; thereafter medical certification is required (➔ p. 95). In many cases, employers have additional schemes that are more generous than SSP. People ineligible for SSP may be eligible for Employment and Support Allowance or Maternity Allowance (➔ p. 765).

Employment and Support Allowance (ESA) can be claimed by people aged ≥16y and under state pension age who are: not in full time education, unable to work due to illness or disability, and unable to claim SSP. There are 2 types of ESA:

- Contributory—for people who have paid sufficient NI contributions. Payable for 1y
- Income-related—for people unable to work for >1y or who have not paid enough NI contributions, and who are not claiming Universal Credit

Payments do not start until 3d into illness. Claims are made by telephone (☎ 0800 055 6688). Claimants can self-certify that they are unfit to work for the first 7d; thereafter medical certification is required (➔ p. 94) until WCA takes place (usually in <13wk). Payments vary according to:

- Age—whether <25y or ≥25y
- Duration of claim—lower rate during the 'Assessment phase' (3d–14wk)
- WCA outcome (➔ p. 94)—lower rate is paid for people placed in the 'work-related activity group' than for those placed in the 'support group'

❗ People claiming ESA may be able to undertake limited work either as part of a treatment programme or if limited hours (<16h/wk) and weekly income from work is less than a specified amount.

Universal Credit ➔ p. 104 *Maternity benefits* ➔ p. 765

Benefits for those with long-term disability

Disability Living Allowance (DLA) Can be claimed for children <16y who fulfil specified UK residence qualifications, and have a disability or illness resulting in mobility problems and/or a need for more care than would usually be expected for a child of that age. Unless terminally ill (when claims are made under 'Special Rules'), the disability/illness must have been present >3mo and be expected to continue for >6mo. Claims forms can be requested by telephone (☎ 0800 121 4600). There are 2 components:

- Mobility component—for children needing help to get around outdoors; two levels and age restrictions apply
- Care component—for children needing help with personal care; three levels. If terminal illness, highest rate is automatically awarded

Personal Independence Payment (PIP) Can be claimed by people aged 16–64y who fulfil specified UK residential requirements, and have a disability requiring assistance that has been present >3mo and is expected to last >9mo (unless terminally ill, when 'Special Rules' apply), regardless of ability to work. Claim forms can be requested by telephone (☎ 0800 917 2222). There are 2 payment components assessed against standard criteria: daily living (activities 1–9) and mobility (activities 10–11):

- Daily living component Paid at standard rate if ≥8 points from activities 1–9, and at enhanced rate if ≥12 points. Enhanced rate is paid automatically if the person has a terminal illness
- Mobility component Paid at standard rate if ≥8 points from activities 10–11, and at enhanced rate if ≥12 points

Attendance Allowance (AA) Can be claimed by people aged ≥65y who are UK residents, and have a disability requiring assistance that has been present >3mo and is expected to last >6mo more (unless terminally ill when 'Special Rules' apply). Claim forms can be requested by telephone (☎ 0800 731 0122). A higher rate is payable if 24h care is required.

Special Rules If a person has a terminal illness and is not expected to live >6mo, claims for DLA, PIP, or AA can be made under 'Special Rules'. This means that the claim is processed much faster and the highest care component rate is automatically awarded. To claim, applicants must complete a standard application form, but this is supported by a medical statement (DS1500 form) completed by a hospital specialist or GP.

Carer's Allowance Can be claimed by people aged ≥16y, on low income and not in full time education, who spend ≥35h/wk caring for a person with a disability who is receiving AA, Constant Attendance Allowance, enhanced rate of the daily living component of PIP, or the middle- or higher-rate care component of DLA. Other benefits (e.g. state pension) may affect eligibility. Claims can be made online via 🖱 www.gov.uk/carers-allowance.

Council Tax reduction and local authority grants People with disability may be eligible for Council Tax reductions and/or certain grants to help them to live in the community—contact local council.

Industrial injury benefits ➔ p. 91

Armed Forces Compensation Scheme ➔ p. 107

Further information

AA 🖱 www.gov.uk/attendance-allowance
Citizen's Advice 🖱 www.adviceguide.org.uk
DLA 🖱 www.gov.uk/disability-living-allowance-children
ESA 🖱 www.gov.uk/employment-support-allowance
PIP 🖱 www.gov.uk/pip
SSP 🖱 www.gov.uk/statutory-sick-pay

Chapter 5

Medicines and prescribing

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NHS prescriptions

> '*A doctor is a man who writes prescriptions till the patient either dies or is cured by nature*'
>
> John Taylor (1694–1761)

At any one time, 43% of adult ♂ and 50% of adult ♀ are taking a prescription medication. More than 1 in 5 regularly take >3 prescription medications; this increases with age and >70% of those aged ≥75y are taking ≥3 regular prescribed medications. In England alone, >2.7 million NHS prescription items are dispensed every day—an average of 18.7 items/head of population/y.

Prescribing forms a major part of any GP's workload. Bad prescribing wastes resources, deprives patients from a chance to benefit, and may cause illness. Medicines should be prescribed only when necessary, and, in all cases, benefits of prescribing should be weighed against risks.

Prescription pre-payment certificate (PPC) For patients who live in England, if not entitled to free prescriptions (Table 5.1) but needing a lot of medication (>3 prescriptions/3mo or >13/y), it is cheaper for a patient to purchase a 'pre-payment certificate'.

There are 3 ways to purchase a PPC:
- Internet 🖱 www.nhsbsa.nhs.uk/HealthCosts
- Telephone ☎ 0300 330 1341
- From a pharmacy registered to sell PPCs. List available at 🖱 https://www.nhsbsa.nhs.uk/help-nhs-prescription-costs/prescription-prepayment-certificates-ppcs

❗ Currently, only patients in England pay prescription charges. There are no prescription charges in Wales, Northern Ireland, or Scotland.

Refunds Send the NHS Business Services Authority a letter explaining the reason for refund to: NHS Help with Health Costs, Bridge House, 152 Pilgrim Street, Newcastle-upon-Tyne, NE1 6SN.
- Full refund—may be claimed if <1mo after purchase the holder becomes entitled to free prescriptions or dies
- Partial refund—may be claimed if the holder dies >1mo after issue or if the holder becomes entitled to free prescriptions 1–4mo after issue

Reclaiming money To reclaim money spent while awaiting an exemption certificate or PPC, ask for an official receipt at the pharmacy when the drug is paid for (FP57 England). Claim money back from any NHS pharmacy with proof of exemption or PPC within 3mo.

Drugs cheaper over the counter (OTC) Many drugs commonly prescribed in primary care (e.g. paracetamol, topical steroid nasal sprays, oral antihistamines, ibuprofen) are cheaper than a prescription charge to buy OTC. Before prescribing for patients who pay prescription charges always consider:
- Is this medication available to purchase OTC?
- Would it be cheaper to buy OTC in the quantities required?

In a bid to ↓ prescribing costs, many PCOs in the UK have directed GPs *not* to supply drugs to patients via NHS prescription when easily available to purchase OTC.

Table 5.1 Free prescription entitlement in England

Circumstances in which free prescriptions can be claimed	How to claim?
Automatic entitlements - Prescription for contraceptive - >60y or <16y of age or age 16–18y in full-time education - Patient or family receiving: Income Support, Income-based JSA, Income-related ESA, or Pension Guarantee Credit. Some people claiming Universal Credit also qualify	Tick the relevant box on the back of the prescription form
Maternity exemption (MatEx) Pregnant women and women who have had a baby <12mo ago	Fill in an exemption application form (FW8) as soon as pregnancy is confirmed Exemption certificates last 1y from estimated date of delivery, or if completed after the baby is born, for 12mo after the baby's date of birth
Certain medical conditions (MedEx) - DM (unless diet-controlled only) - Myxoedema or need for thyroxine - Hypoparathyroidism - Epilepsy requiring continuous anticonvulsants - Permanent fistula (e.g. colostomy) needing stoma dressing/appliance - Hypoadrenalism (including Addison's disease) and on replacement therapy - Hypopituitarism, including diabetes insipidus - Myasthenia gravis - Cancer - Unable to go out without the help of another person due to a continuing physical disability	Fill in an exemption application form (FP92A) as soon as the condition is confirmed Requires a doctor's signature to confirm the condition. Certificates last 5y or until 60th birthday, if sooner
Tax credit exemption People who have low income and receive either CTC, or WTC + CTC, or WTC + disability element	NHS tax credit exemption certificate is received automatically at the time tax credits are applied for
Low-income scheme Low income and the individual/family has assets of <£16,000 in property and/or savings	Complete form HC1 available to order or download from the NHS Business Services Authority website (www.nhsbsa.nhs.uk) or order from ☎ 0345 603 1108 HC2 certificates (full help with health costs) entitle the holder to free prescriptions
War pensioners—prescriptions related to pensionable condition only	Contact Veterans UK ☎ 0808 1914 218

! Leaflet HC11, *Help with Health Costs*, is available from the NHS Business Services Authority. Information is also available on their website in a variety of different languages. www.nhsbsa.nhs.uk

Writing prescriptions

⚠ Legal responsibility for writing prescriptions lies with the person who signs the prescription form.

British National Formulary (BNF) Contains a list of all drugs that a registered medical practitioner can prescribe on NHS prescription. It does not include homeopathic drugs, nor aids and appliances. There is a separate but linked *BNF* concerned with prescribing for children. Dentists and nurses have their own limited formulary. *Further information*: 🖱 https://bnf.nice.org.uk/

Claiming for items dispensed by a non-dispensing GP All GPs may claim payment for dispensing certain items that are supplied and personally administered by the GP or practice staff on behalf of the GP. Claims are made on form FP10 (GP10) to the NHS Business Services Authority and must state the name of the patient, item dispensed, and manufacturer of the item. Claimable items include:

- Vaccines
- Anaesthetics
- Injections
- Sutures
- Skin closing strips
- IUCDs
- Contraceptive caps/diaphragms
- Diagnostic reagents
- Pessaries that are appliances (e.g. ring pessary)

❗ Different arrangements apply for high-volume vaccines, e.g. influenza.

Prescription writing NHS prescriptions are written on form FP10 (GP10 in Scotland). They should be legible and in indelible ink. They are valid for 13wk from the date written on them. Include:

- Patient details—full name, address, and age/date of birth if <12y
- Date
- Full name of the drug (not abbreviated), with quantity to be supplied and dose interval (avoid the use of decimal points, e.g. for quantities <1g, write in mg). If you want a description of the drug included on the label, then write it on the prescription (e.g. 'for asthma')
- Deletion of any unused space (e.g. by striking through)
- Name and address of the prescriber
- Must be signed in ink by an authorized prescriber

❗ Special rules apply for controlled drugs—➔ p. 125.

Computer-issued prescriptions (form FP10(C)) Should contain the same information as their handwritten equivalents.

Electronic Prescription Service (EPS) Removes the need for paper prescriptions. The prescription is generated by the prescriber in the same way as a computer-issued prescription but the prescriber's electronic signature is added (using the user's NHS Smartcard and password) and the prescription is sent electronically directly to a pharmacy of the patient's choice, where it is dispensed in the normal way.

Prescribing for people going abroad

- Do not provide NHS prescriptions for conditions that might arise while a patient is away, e.g. traveller's diarrhoea

- Prescribing interval for repeat medication should be related to the next time that medication would normally be reviewed (generally <13wk). The prescriber retains medicolegal responsibility for the duration of the prescription. If a prescription is issued for the patient's stay abroad (e.g. if repeat supplies cannot be obtained at the destination or narrow therapeutic index), warn the patient to consult a doctor for regular monitoring or for unforeseen medical problems while away

Non-NHS prescriptions The same rules apply to the writing of private prescriptions as NHS prescriptions but private prescriptions should not be written on FP10 forms. Hand-written private prescriptions are usually written on headed notepaper. Computer-generated private prescriptions are often printed on the counterfoil of the FP10 form. Electronically transmitted prescriptions can be electronically marked as 'private prescriptions'.

Private prescriptions for controlled drugs Controlled drugs in Schedules 2 and 3 (including temazepam) presented for dispensing in the community (but not in hospitals) must be written on specially designated forms available from local PCOs. These forms must include the prescriber's unique 6-digit identification number issued for their private prescribing activity.

Dentists (*BNF*—Appendix DPF) Can prescribe medication for dental conditions to their NHS patients on form FP10(D) (GP14 in Scotland).

Other healthcare practitioners with prescribing rights A wide range of healthcare practitioners can undertake supplementary training to become independent prescribers, including nurses, paramedics, community pharmacists, optometrists, physiotherapists, and podiatrists.

Emergency supply of medicines by pharmacists In emergency situations any pharmacist can dispense prescription-only medicines (POM). In general ≤5d supply can be dispensed.

Patient information All newly licensed/relicensed medicines dispensed in an original pack must be accompanied by a patient information leaflet (PiL). Even though most drugs are now supplied with PiLs, prescribers should make patients aware of 'substantial or special risks'. Information regarded as important by patients includes: name of drug; what to do if a dose is missed; purpose of treatment; precautions (e.g. effect on driving); when/how to take the medicine; problems with alcohol/other drugs; unwanted effects and what to do about them.

Security of prescriptions NHS prescription fraud is common and wastes valuable NHS resources. GP surgeries have a responsibility to:

- Keep paper prescriptions secure
- Ensure that Smartcards are not left unattended or passwords shared
- Be alert to irregularities associated with prescriptions/prescribing, e.g. requests for repeat prescriptions not ordered by patients or excessive quantities of drugs, altered paper prescriptions, patients who have multiply registered to obtain prescription drugs

Report suspected fraudulent use of NHS prescriptions to the NHS Counter Fraud Authority: ☎ 0800 0284060 🖱 www.cfa.nhs.uk/reportfraud

Medicines management

Defined as 'facilitating the maximum benefit and minimum risk for medicines for individual patients'. Encompasses the way medicines are selected, procured, delivered, prescribed, administered, and reviewed.

Generic prescribing Use of generic, rather than brand name, when prescribing is one of the simplest ways to ↓ cost of drugs to the NHS without compromising patient care. As long as the drug's patent is valid, the company that developed the drug will derive income from prescription whatever the name on the prescription. Once the patent has expired, competitors can manufacture the drug and market it under its generic or an alternative brand name. If prescribed generically, the pharmacist decides which brand to supply and market forces drive price ↓.

Reasons not to prescribe generically

- Drugs with a low therapeutic index e.g. lithium, carbamazepine, phenytoin, ciclosporin—small differences in plasma concentrations can be clinically significant
- Modified-release formulations e.g. diltiazem, or theophylline products. Composition/pharmacokinetic properties are difficult to standardize
- Formulations containing ≥2 drugs Some do have generic names (e.g. co-amilofruse 5/40, co-codamol)—others do not. Do not make up a generic name if the combination drug does not have one

Practice and locality formularies An agreed practice or locality formulary is an effective way to promote best practice in prescribing, and limit costs. Those compiling formularies should consider: evidence of efficacy; safety; cost-effectiveness; national guidelines and local policies.

Concordance A process of prescribing and medicine-taking based on partnership. Patient concordance (or rather lack of it) is a major challenge in general practice. For drugs to be optimally effective they should be taken as directed by the prescriber. Concordance sufficient to attain therapeutic objectives occurs about 50% of the time—1 in 6 patients take medication exactly as directed; 1 in 3 take medication as directed 80–90% of the time; 1 in 3 take medication as directed 40–80% of the time; the remaining 16–17% take medication as directed <40% of the time.

'White-coat concordance' Phenomenon in which 90% of patients take regular medication as directed for a period before a check-up—may mask effects of non-concordance.

Consequences of non-concordance

- Failure to attain therapeutic targets, e.g. not taking antihypertensive medication, results in increased risk of stroke
- Wastage of precious resources. ~£250 million worth of medicines are returned to pharmacies each year for disposal—the true quantity wasted is many times that

Ways to improve concordance

- Use simple language and avoid medical terms; discuss reasons for treatment and consequences of not treating in terms the patient can understand; seek the patient's views on his/her condition and agree a course of action before prescribing

- Explain what the drug is, its function, and (if known and not too complex) its mechanism of action
- Keep the drug regimen as simple as possible—od or bd dosing is preferable, especially long term; discuss how the patient will manage the regimen within his/her daily schedule and try to tie in with daily routine (e.g. take one in the morning when you get up). Give clear verbal instructions and reinforce with written instructions if a complex regimen, the patient is elderly, or the understanding of the patient is in doubt
- Discuss possible side effects
- Respond to any questions the patient has
- If necessary arrange review within a short time of starting medicine to discuss progress or queries, or arrange follow-up by another member of the primary healthcare team (e.g. asthma nurse to check inhaler technique 2–3wk after starting inhaler)
- Monitor repeat prescriptions

Medication errors The PRACtICe Study (2012) showed that 1 in 20 prescriptions issued in general practice contain an error: 42% are minor, 54% moderate, and 4% severe. Common errors include incomplete information on prescriptions; dose, strength, or dose timing errors; and monitoring errors (where necessary monitoring was not requested).

Minimizing medication errors

- Prescriber actions Read aloud prescriptions to patients to check for errors and ensure patient understanding; if unfamiliar with a drug initiated in secondary care, clarify before prescribing; review patients on new drugs after <6wk and do not put medication on repeat until stable; confirm important information (e.g. drug allergies) with patients even if you know them well; become familiar with prescribing safety features on practice IT systems and use them
- Practice actions Appoint a prescribing lead; use a practice/locality formulary; discuss adverse prescribing events and learn from mistakes; update medical records as soon as possible after notification of medication alterations by other services; use call–recall systems to ensure regular monitoring tests are done and results available before prescription requests are due; train dedicated staff to manage prescription requests; highlight queries to GPs; ensure 'difficult' patients (e.g. those on many drugs) have a named GP to manage their medication
- IT solutions Use alerts, warnings, and other safety features built into practice computer software to highlight potential pitfalls; perform regular prescribing audits for high-risk situations, e.g. drug monitoring for potentially dangerous drugs such as methotrexate

Further information

Avery T, et al. (2012) The PRACtICe Study. 🖱 http://www.gmc-uk.org/about/research/12996.asp

GMC (2013) Good practice in prescribing and managing medicines and devices. 🖱 www.gmc-uk.org/ethical-guidance/ethical-guidance-for-doctors/prescribing-and-managing-medicines-and-devices

NICE (2009) Medicines adherence. 🖱 https://www.nice.org.uk/guidance/cg76

Repeat prescribing

80% of NHS prescriptions are for repeat medication. Good practice is essential to ensure wastage (>10% of total prescribing costs) is kept to a minimum. *Essential elements are*:

- Written explanation of the repeat prescribing process for patients and carers
- Practice personnel with dedicated responsibility ensure patient recall and regular medication review
- Agreed practice policies for repeat prescriptions, e.g. duration of supply; procedure if someone 'runs out' but is not authorized to have more
- Authorization check each time a prescription is signed
- Compliance check for under- or overuse (prescription frequency)
- Equivalence check that all regular prescriptions are for the same duration of treatment so that prescription requests can be synchronized
- Regular housekeeping keeps records of medication up to date (including dosage instructions)—particular care is needed after hospital discharge when medication could have been substantially changed
- Training of practice staff

Review process Invite the patient ± carer. *Areas to cover*:

- Explain what you want to do in the review and the reasons for it
- Compile a list of all medicines being taken/used including: prescribed medication; OTC drugs; herbal/homeopathic medicines; illicit drugs; and medicines borrowed from others. Compare the list of drugs generated with the prescription record
- Concordance. Find out whether and how medication is taken
- Explore understanding of the purpose of the medication and consequences of not taking it and how much, how often, when
- Discuss misconceptions/queries
- Ask about side effects
- Review relevant monitoring tests, e.g. lithium level; TFTs; INR; HbA1c
- Review practical aspects. Problems ordering/receiving repeat prescriptions; using medicines, e.g. problems opening containers; with formulations, e.g. difficulty swallowing tablets; reading labels—can request large print; remembering to take medication—consider reminder chart, multi-compartment compliance aid, altering times of doses to fit in better with daily schedule
- Check necessity and appropriateness of all prescriptions (Figure 5.1)

Comparative practice data PCO prescribing teams can provide comparative prescribing data for practices within their area. Comparing practice data against that of other local practices can highlight over- or under-prescribing and expensive drugs being used for which there are cheaper, equally effective alternatives.

Further information

BNF 🖱 https://bnf.nice.org.uk/

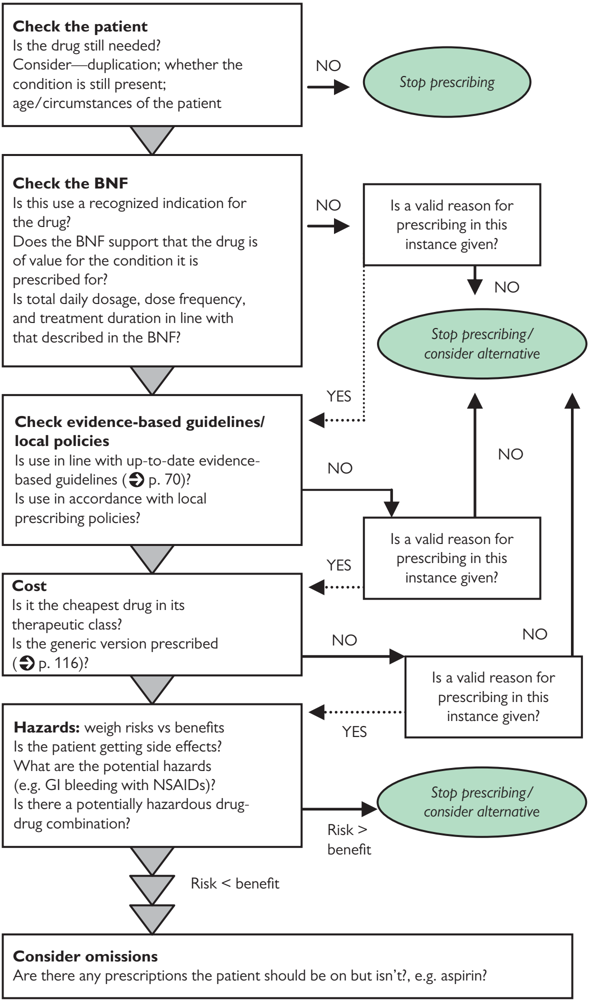
Check the patient
Is the drug still needed?
Consider—duplication; whether the condition is still present; age/circumstances of the patient
NO
Stop prescribing
Check the BNF
Is this use a recognized indication for the drug?
Does the BNF support that the drug is of value for the condition it is prescribed for?
Is total daily dosage, dose frequency, and treatment duration in line with that described in the BNF?
NO
Is a valid reason for prescribing in this instance given?
NO
Stop prescribing/ consider alternative
YES
Check evidence-based guidelines/ local policies
Is use in line with up-to-date evidence-based guidelines (➔ p. 70)?
Is use in accordance with local prescribing policies?
NO
NO
Is a valid reason for prescribing in this instance given?
YES
Cost
Is it the cheapest drug in its therapeutic class?
Is the generic version prescribed (➔ p. 116)?
NO
NO
Is a valid reason for prescribing in this instance given?
YES
Hazards: weigh risks vs benefits
Is the patient getting side effects?
What are the potential hazards (e.g. GI bleeding with NSAIDs)?
Is there a potentially hazardous drug-drug combination?
Stop prescribing/ consider alternative
Risk > benefit
Risk < benefit
Consider omissions
Are there any prescriptions the patient should be on but isn't?, e.g. aspirin?

Figure 5.1 Deciding whether a prescribed drug is appropriate

Adverse drug reactions

> '*I don't want two diseases—one nature-made and one doctor-made*'
>
> Napoleon Bonaparte, St Helena (1820)

An adverse drug reaction is an unwanted or harmful reaction which occurs after administration of a drug or drugs and is suspected or known to be due to the drug(s). Any drug may produce unwanted or unexpected effects. Common side effects are listed in the *BNF* or drug data sheet but any patient can have an allergic reaction or idiosyncratic response to any drug.

Types of adverse reaction

- Type A ('Augmented') drug reactions are related to the normal pharmacological action of the drug. As such, they are common, predictable, and usually dose dependent. Examples include: constipation with opioids; dry mouth with amitriptyline; cough with ACE inhibitors. Usually managed by reducing the dose or withholding the drug
- Type B ('Bizarre') reactions are not related to the usual pharmacological action of the drug. They are uncommon, not predictable, and may not be dose dependent. Examples include: anaphylaxis with penicillin; cholestatic jaundice with co-amoxiclav

Management of suspected adverse drug reactions

- Severe reactions (e.g. anaphylaxis, respiratory depression, Stevens–Johnson syndrome)—take emergency action
- If less severe, assess the patient—take a history of the presenting symptoms and a detailed drug history. Has the patient ever had a similar reaction before? A temporal relationship between starting or ↑ the dose of a drug may suggest a drug reaction. Examine the patient as directed by the history and arrange any tests needed (e.g. blood test if lithium or digoxin toxicity is suspected)
- Check the drug literature—is this a recorded side effect of any implicated medication(s)?
- Consider whether the reaction may be due to another cause, e.g. another medical condition, drug interaction, or OTC medication
- Consider whether to stop the drug or reduce the dose. If the drug is stopped, monitor to see if the adverse reaction resolves. Treat symptoms if needed (e.g. antihistamine for itching)
- Record any adverse drug effects in the patient's medical record
- Consider reporting the adverse reaction via the Yellow Card Scheme

Reporting adverse drug events In the UK, the 'Yellow Card Scheme' allows both healthcare professionals and patients to report adverse drug events to the Medicines and Healthcare products Regulatory Agency (MRHA). Reports can be filed online via 🖱 www.yellowcard.mhra.gov.uk. The scheme covers:

- Adverse reactions associated with medicines and vaccines (including blood products and complementary medicines)
- Adverse incidents associated with medical devices
- Defective or counterfeit medicines
- Side effects or safety concerns with e-cigarettes

~25,000 Yellow Card reports are submitted to the MHRA each year. By collating adverse events, product data sheets can be updated as side effects are recognized and warnings issued and/or drugs withdrawn if serious safety concerns emerge.

Report

- Serious suspected adverse drug reactions to established drugs and vaccines (including OTC, herbal, and unlicensed or off-label medicines)
- All suspected adverse drug reactions (including those not deemed serious) to newly licensed medicines identified by the ▼ symbol in the *BNF*

Prevention of adverse reactions

- Never use a drug unless there is a good indication
- Always ask patients if they have had reactions previously to a drug before prescribing
- Ask about other drugs patients are taking (including self-medication); consider interactions
- Consider the effects of age and hepatic or renal impairment
- Prescribe as few drugs as possible—the more drugs, the more likelihood of interactions
- Give clear instructions about how to take the drug
- Wherever possible use drugs you are familiar with; if using a new drug, be alert to side effects
- Warn patients about potentially serious side effects (e.g. risk of GI bleeding with NSAIDs)

Defective medicines A medicine which does not conform to its specification is deemed defective. Report suspected defective medicines, with as much detail as possible via the Yellow Card Scheme.

Consumer Protection Act (1987) If a patient is damaged by a defective product, liability falls on the producer unless outside the EC when it falls on the importer. If the importer cannot be identified, liability falls on the supplier. This is important for GPs. Those who dispense are at greatest risk but all GPs occasionally supply drugs in an emergency or for procedures within the surgery (vaccinations, minor surgery, contraception). Always record manufacturer, batch number, and expiry date when using such drugs and keep records of storage of drugs and maintenance of equipment.

Sources of drug information

- *British National Formulary* (BNF) 🖱 https://bnf.nice.org.uk/
- Electronic Medicines Compendium 🖱 www.medicines.org.uk
- Interactive Drug Analysis Profiles (iDAPs)—summary of adverse events reported to the MHRA for individual drugs 🖱 https://yellowcard.mhra.gov.uk/idap
- Regional and district medicine information services

Licensing of medicines

In the UK, the Medicines Act (1968) makes it essential for anyone who manufactures or markets a drug for which therapeutic claims are made to hold a licence. The Licensing Authority, working through the Medicines and Healthcare products Regulatory Agency (MHRA), can grant both Manufacturer's Licence and Marketing Authorization (which allows a company to market and supply a product for specified indications). Although doctors usually prescribe according to the licensed indications, they are not obliged to.

Unlicensed and off-label drugs Unlicensed drugs are drugs that do not have a product licence for use in the UK. 'Off-label' drugs are drugs that have a product licence but are being used for indications not covered by that product licence.

Prescribing outside licence There may be occasions when a doctor feels it is necessary to prescribe outside a drug's licence.

- Generic formulations for which indications are not described. The prescriber has to assume the indications are the same as for branded formulations
- Use of well-established licensed drugs for proven indications not covered by the product licence, e.g. amitriptyline for neuropathic pain
- Use of drugs for conditions where there are no other treatments, even if the evidence of their effectiveness is not well proven. This often occurs in secondary care when new treatments become accepted. GPs may become involved if a patient is discharged to the community and the GP is asked to continue prescribing. ❗ The person signing the prescription is legally responsible
- Use of drugs for individuals not covered by their licensed indications—frequently occurs in paediatrics

⚠ Before prescribing any medication (whether within or outside the licence) weigh risks against benefits. The more dangerous the medicine and the flimsier the evidence base for treatment, the more difficult it is to justify the decision to prescribe.

When prescribing licensed drugs for unlicensed indications, it is important to inform patients and carers of what you are doing and why. Explain that the patient information leaflet (PiL) will not have information about the use of the drug in these circumstances. Record in the patient's notes your reasons for prescribing outside the licensed indications for the drug.

Clinical trials Drug discovery and development is a protracted process (>10y) costing huge sums of money (~£100 million). Clinical testing is conventionally divided into five stages:

- Phase I trials Clinical pharmacology in normal volunteers
- Phase II trials Preliminary small-scale studies
- Phase III trials Large-scale trials (several thousands of patients often). Once complete, application is made for a licence to sell the drug

- Phase IV trials Post-marketing surveillance—large-scale follow-up of patients using the drug to establish evidence of long-term efficacy and safety
- Phase V trials Further trials to compare efficacy and safety with other marketed compounds and explore new indications

GPs are unlikely to be involved before phase III. Taking part in trials can benefit both patients and the practice but consider proposals carefully before embarking on a project.

Research in general practice ➔ p. 55

Questions to ask before agreeing to take part in a clinical trial

- Are the aims and objectives of the study defined?
- What is the design?
- Which drug is to be tested?
- What are the end points?
- Are the criteria for identifying patients clear and explicit?
- Are the numbers to be recruited specified and feasible?
- Are the observations to be made clearly and vigorously defined?
- Are the arrangements for providing information to patients and for obtaining informed consent satisfactory?
- Has ethical approval of the study been obtained?
- Are the financial arrangements clearly set out (minimum—reimbursement of patients' expenses and reimbursement of practice expenses)?
- Has adequate provision been made for compensation in the event of injury to patients in the course of the study?

Controlled drugs

Misuse of Drugs Act (1971) Controls manufacture, supply, and possession of controlled drugs (CDs). Penalties for offences are graded according to perceived harmfulness of the drug into three classes:

- Class A—e.g. cocaine, diamorphine (heroin), methadone, LSD, ecstasy
- Class B—e.g. oral amphetamines, barbiturates, cannabis, codeine
- Class C—e.g. most benzodiazepines, tramadol and anabolic steroids

Misuse of Drugs Regulations (2001) Defines persons authorized to supply and possess CDs while carrying out their professions and describes the way this is to be done. Five schedules of drug are defined:

- Schedule 1 Drugs not used for medicinal purposes, e.g. LSD. Possession and supply are prohibited except with special licence
- Schedule 2 Drugs subject to full CD controls (written dispensing record, kept in locked container, CD prescription regulations), e.g. diamorphine, cocaine, pethidine
- Schedule 3 Partial CD controls (as Schedule 2, but no need to keep a register—some drugs subject to safe custody regulations), e.g. barbiturates, temazepam, midazolam, tramadol, meprobamate, buprenorphine
- Schedules 4 and 5 Most benzodiazepines, anabolic and androgenic steroids, hCG, growth hormone, codeine. CD prescription requirements do not apply nor do safe custody requirements

❗ Controlled drugs are identified throughout the *BNF* by the box symbol containing the letters CD (controlled drug) together with a number (i.e. CD1 or CD4-1). The number refers to the Schedule of the Misuse of Drugs Regulations (2001) that the drug is classified within.

Controlled drugs register All healthcare professionals who hold personal stock of any Schedule 2 drugs must keep their own controlled drugs register, and they are personally responsible for keeping this accurate and up to date. Out-of-date drugs should be recorded and destroyed in the presence of an authorized witness (police, PCO official).

Prescriber's responsibilities If prescribing controlled drugs for medicinal purposes, you have a responsibility:

- To avoid creating dependence by unnecessarily introducing controlled drugs to patients—➔ p. 168 and ➔ p. 183
- For careful monitoring to ensure the patient does not gradually ↑ the dose of drug to a point where dependence becomes more likely
- To avoid being an unwitting source of supply for addicts. If you suspect an addict is going round surgeries with intent to obtain supplies, contact your PCO so that they can issue a warning to other practices and/or the NHS Counter Fraud Authority. ☎ 0800 0284060 🖱 www.cfa.nhs.uk/reportfraud

Prescribing for drug misusers ➔ p. 163

Notification of drug misusers ➔ p. 163

Writing prescriptions for CDs Any prescription for Schedule 2 and 3 controlled drugs (with the exception of temazepam) must contain the following details written so as to be indelible:

- The patient's full name, address, and age—if the patient is homeless, 'no fixed abode' is an acceptable address
- The patient's NHS (in Scotland, Community Health Index) number
- Name and form of the drug, even if only one form exists
- Strength of the preparation and dose to be taken
- The total quantity of the preparation, or the number of dose units, to be supplied in both words and figures, e.g. 'Morphine sulfate 10mg (ten milligram) tablets, one to be taken twice daily. Supply 60 (sixty) tablets, total 600 (six hundred) milligrams'
- Signature of the prescriber (currently must be handwritten, although electronic signatures for EPS prescriptions are likely to be approved in the near future) and date. It is good practice to include the GMC number of the prescriber as well
- The address of the prescriber

❗ Apart from in exceptional circumstances, prescriptions for CDs in Schedules 2, 3, and 4 should be limited to a supply of ≤30d treatment. The validity period of NHS and private prescriptions for Schedules 1, 2, 3, and 4 CDs is restricted to 28d. Schedules 2 and 3 drugs should not be prescribed on repeat prescriptions or under repeat dispensing schemes.

Travelling abroad with controlled drugs For patients or doctors travelling abroad with Schedule 2 or 3 drugs, an export licence may be required. Further details are available from 🖱 www.gov.uk/travelling-controlled-drugs. Patient applications to the Home Office for an import/export licence for a CD must be accompanied by a supporting letter from the prescribing doctor stating the:

- Patient's name and address
- Quantities of drugs to be carried
- Strength and form in which the drugs will be dispensed
- Country of destination
- Dates of travel to and from the UK

For clearance to import the drug into the country of destination, it is advisable to contact the Embassy or High Commission of that country prior to departure

Further information

Advisory Council on the Misuse of Drugs 🖱 www.gov.uk/government/organisations/advisory-council-on-the-misuse-of-drugs

BNF Controlled drugs and drug dependence. 🖱 https://bnf.nice.org.uk/guidance/controlled-drugs-and-drug-dependence.html

Faculty of Pain Medicine Opioids aware: a resource for patients and healthcare professionals to support prescribing of opioid medicines for pain 🖱 https://www.rcoa.ac.uk/faculty-of-pain-medicine/opioids-aware

NICE Controlled drugs: safe use and management 🖱 www.nice.org.uk/guidance/ng46

Prescribing for special groups

Borderline substances (*BNF* Appendix 2) For certain conditions, foods and toilet products can be regarded as drugs and prescribed via NHS prescription (e.g. gluten-free foods for coeliac disease, nutritional supplements for disease-related malnutrition). The Advisory Committee on Borderline Substances advises on which products are available for certain specified conditions. Products should not be prescribed for any other condition. Use form FP10 (GP10 in Scotland) and endorse with the letters 'ACBS'.

Renal impairment

Degree of renal impairment Estimated glomerular filtration rate (eGFR), based on serum creatinine, age, gender, and ethnic origin, is now provided to GPs when renal function tests are done.

- Mild renal impairment eGFR 60–89mL/min/1.73m^2
- Moderate renal impairment eGFR 30–59mL/min/1.73m^2
- Severe renal impairment eGFR <30mL/min/1.73m^2

For some drugs (e.g. DOACs) dosage recommendations are based on calculation of creatinine clearance. This can be calculated as follows:

[(140 − age in years) × weight in kg] ÷ [72 × serum creatinine in mg/dL]

Creatinine clearance can only be calculated if the creatinine level is stable. For ♀, the result is multiplied by 0.85.

! Renal function ↓ with age but may not be reflected by raised creatinine due to ↓ muscle mass. Always assume mild to moderate renal failure if prescribing for the elderly.

Drug effects of impaired renal function

- Inability to excrete the drug—may cause toxicity. Dose reduction or increase in interval between doses may be necessary
- Increased sensitivity to drugs—even if elimination is unimpaired
- Poor tolerance of side effects—nephrotoxic drugs in particular may have more serious side effects
- Lack of effectiveness when renal function is reduced

! For patients on dialysis, consult your local renal unit if unsure.

Hepatic impairment Problems do not tend to arise until late stages of liver failure when there is jaundice, ascites, or evidence of encephalopathy. Problems are due to:

- Impaired drug metabolism—many drugs are metabolized by the liver. In severe liver failure, dose may need to be ↓ ± dosage interval ↑. A few drugs are excreted in the bile unchanged and may accumulate in patients with obstructive jaundice (e.g. rifampicin, fusidic acid)
- Hypoproteinaemia—liver failure is associated with ↓ plasma protein. This affects binding of drugs. Highly protein-bound drugs (e.g. phenytoin, prednisolone) can become toxic in normal dosage
- Hepatotoxicity—any liver toxicity of drugs (e.g. diclofenac) will have ↑ effect if hepatic reserve is already ↓
- Clotting—blood clotting factors are made in the liver. In liver disease, effects of oral anticoagulants are ↑
- Encephalopathy—drugs that depress cerebral function (e.g. benzodiazepines, opioids) can precipitate encephalopathy

- Fluid retention—drugs causing fluid retention (e.g. NSAIDs) make oedema and ascites worse

Palliative care ➲ p. 1011 **Drugs and sport** ➲ p. 475

Prescribing for the elderly ➲ p. 194

Pregnancy ➲ p. 764. Drugs taken by the mother can harm the fetus at any stage in pregnancy. *Mechanisms*:

- 1st trimester—teratogenesis causing congenital malformations. Greatest risk is from 3–12wk gestation
- 2nd/3rd trimesters—toxic effects; effects on growth/development
- Around labour—may affect labour or have adverse effects on the newborn baby

Only prescribe if essential, especially in the 1st trimester. Stick to tried and tested drugs when possible; use smallest effective dose; avoid new drugs. ❗ Lack of information does not imply safety.

Breastfeeding Drugs taken by a breastfeeding mother can affect the child by inhibiting lactation or entering the milk and causing toxicity to the infant. Therapeutic doses in the mother can cause toxicity in the infant if the drug is concentrated in milk (e.g. iodides). Avoid prescribing, wherever possible and stick to tried and tested drugs.

Driving while taking drugs ➲ p. 99

Prescribing for children Keep all medicines out of the reach of children (and, preferably, in a locked cupboard). Dispose of unwanted medicines by returning them to a pharmacy for destruction.

- Children differ from adults in their response to drugs. Consult the *BNF for Children* or before prescribing unfamiliar drugs. Always check doses carefully. Many drugs commonly used for adults are not licensed for use with children
- Paediatric suspensions may contain sugar. For long-term use or children having frequent prescriptions, consider sugar-free versions
- Do not advise adding medicines to infant feeding bottles—they may interact with milk and the dose will be ↓ if not all contents are drunk
- Report serious adverse reactions and adverse reactions to new drugs marked in the *BNF for Children* with a ▼ on Yellow Cards ➲ p. 120.

Further information

BNF 🖱 https://bnf.nice.org.uk/guidance/

- Prescribing in the elderly
- Prescribing in pregnancy
- Prescribing in palliative care
- Prescribing in children
- Prescribing in hepatic impairment
- Prescribing in renal impairment
- Prescribing in breastfeeding
- Drugs and sport

BNF for Children 🖱 https://bnfc.nice.org.uk/
Electronic Medicines Compendium 🖱 www.medicines.org.uk
UK Teratology Information Service ☎ 0344 892 0909 🖱 www.uktis.org

Non-drug interventions and complementary medicine

Across the world there has been a shift away from the traditional 'pill-for-every-ill' approach to medicine. Antibiotic resistance and frequent press stories about adverse side effects of medication have led to patients seeking alternative ways to manage their medical problems. In the UK, ~90% of the population have tried complementary or alternative medicine (CAM) at some time (Table 5.2). But, although CAM undoubtedly helps many individuals, its use remains controversial.

Reasons for caution Lack of:

- **Evidence of effectiveness** Although almost half of all clinical trials examine the effectiveness of non-drug and complementary therapies, unlike drug information, until recently access to independent appraisals of effectiveness of these treatment has not been easily accessible to GPs
- **Regulation of practitioners** Some practitioners offering non-drug and/or complementary treatments have professional registration (e.g. physiotherapists, osteopaths, and chiropractors). However, at present, anyone can set up as a practitioner of alternative medicine. Therefore, it is important to stress to patients contemplating CAM that they should find a reputable practitioner with accredited training who is a member of a recognized professional body and carries professional indemnity insurance
- **Regulation of products** At present, most complementary 'medicines' are sold as foods rather than medicines and do not hold a product licence. No licensing authority has assessed efficacy, safety, or quality, and interactions with conventional medicines are unknown. Complementary medicines can, and do, cause adverse effects—just because they are 'natural' does not mean they are safe

Legal position of GPs practising CAM Conventionally trained doctors can administer alternative treatments. The 'Bolam test' applies—in other words, if a doctor has undergone appropriate training and practises in a way that is reasonable and would be considered acceptable by a number of other medically qualified complementary practitioners, his or her actions are defensible.

Handbook for Non-drug Interventions (HANDI) Launched by the Royal Australian College of GPs in 2015 to promote effective non-drug treatments, making them visible and easy to use. HANDI is an online formulary of non-drug interventions for use in primary care, which have solid evidence of their effectiveness. Based on the idea of modern pharmacopoeias, each HANDI entry includes indications, contraindications, side effects, and 'dosing'. Some interventions require referral to other practitioners, e.g. physiotherapists. Others (e.g. use of auto-inflation for glue ear) can be administered directly by GPs. The HANDI project is freely accessible via 🖱 www.racgp.org.au/handi

Table 5.2 Commonly used complementary therapies

Therapy	Features
Acupuncture	Needles are used to alleviate symptoms or cure disease. Mechanism of action remains unclear. Two broad forms: • *Traditional acupuncture* Based on Chinese medicine where health is a balance between ying and yang. Illness is imbalance, and treatment aims to restore balance • *Modern acupuncture* Uses modern anatomy and physiology **Evidence** Mixed—good evidence for back pain, idiopathic headache, and migraine; knee pain; and postoperative nausea/vomiting **Variants** Auriculotherapy; transcutaneous electrical nerve stimulation (TENS); reflexology
Homeopathy	From the Greek, meaning 'treatment of similars'. Works on the principle that like cures like. A remedy is chosen that mimics symptoms displayed by the patient. Most remedies are serially diluted in steps of 1:10 (decimal x) or 1:100 (centesimal c). Manufacture is controlled by the Medicines Act **Evidence** With higher dilutions (>12c), a theoretical problem arises as the solution may not contain any molecules of the mother substance
Herbal medicine	Use of plants for medicinal purposes. Although many traditional medicines were developed from herbal sources (e.g. digoxin, morphine), herbal medicines use plant extracts and not isolated constituents. All herbal medicines must be registered to be sold in the UK **Evidence** Trial evidence supports use of many herbal remedies, including: saw palmetto (benign prostatic hyperplasia); echinacea (common cold); St John's wort (depression); feverfew (migraine) **Variants** Aromatherapy is the use of concentrated aromatic plant oils. Oils commonly used include: lavender (insomnia, burns, blisters); tea tree (skin infection, head lice); peppermint (indigestion); valerian (anxiety, insomnia)
Dietary manipulation	**Healing foods** Are commonly used, e.g. cranberry juice for UTI; soya to ↓ menopausal symptoms; ginger to ↓ nausea **Nutritional medicine** Involves giving supplements of vitamins, minerals, amino acids, or essential fatty acids. There is some evidence of effectiveness, e.g. glucosamine for OA; calcium and vitamin D supplements to ↓ osteoporosis risk; vitamin B_6 for PMT **Probiotics** Orally administered microbial cell preparations. Some evidence of effectiveness for prevent of diarrhoea for patients taking antibiotics, and treatment of GI conditions **Environmental medicine** Based on the premise that individuals develop intolerances to environmental substances—most commonly foods. Exclusion diets improve symptoms. May be effective treatment for IBS and migraine. Most common culprits are caffeine, milk, gluten, and citrus fruit
Osteopathy and chiropractic	Physical treatments aimed at restoring the alignment of joints and improving functioning of the body. Already under statutory regulation. Good evidence of effectiveness, particularly for back pain
Hypnotherapy	Consists of training the patient to relax very deeply—often with a focus, a scene, smell, touch sensation, or colour to aid this process. Frequently used for smoking cessation. May be helpful for pain relief

Chapter 6

Minor surgery

'A minor operation: one performed on someone else'

Penguin Dictionary of Humorous Quotations (2001)

Providing minor surgery *132*
Basic techniques *134*
Removal of skin lesions *136*
Commonly performed injections *138*

Providing minor surgery

Minor surgery Under the GMS contract, minor surgery can be provided as an additional service or directed enhanced service. Arrangements for minor surgery may be different for practices with other contractual arrangements.

Minor surgery as an additional service

- Includes curettage and cautery and, in relation to warts, verrucae, and other skin lesions, cryocautery
- In all cases a record of consent of the patient to treatment and a record of the procedure itself should be kept
- Payment is included within the global sum payment. If a practice does not want to provide this service, it must 'opt out' and global sum payment is ↓ by 0.6%

Minor surgery as a directed enhanced service Extends the range of procedures beyond those practices are expected to do as an additional service. For purposes of payment, procedures are divided into 3 groups:

- Injections—muscles, tendons, bursae, and joints
- Invasive procedures—including incisions and excisions
- Injections of varicose veins and piles

Payment Treatments are priced according to the complexity of the procedure, involvement of other staff, and use of specialized equipment. Terms for this must be negotiated locally.

Qualification to provide minor surgery Practices can provide extended minor surgery (as a directed enhanced service or equivalent) if they can demonstrate they have appropriate facilities and personnel (partner, employee, or subcontractor) with the necessary skills. This includes:

- Adequate equipment
- Premises compliant with national guidelines as contained in 'Health Building Note 46: General Medical Practice Premises' (DH)
- Nursing support
- Compliance with national infection control policies—sterile packs, disposable sterile instruments, etc.
- Ongoing training for involved personnel in minor surgery, related skills, and resuscitation techniques
- Regular audit and peer review to monitor clinical outcomes, rates of infection, and procedure

Location and equipment A suitable room, adequate lighting, the appropriate equipment, and sufficient uninterrupted time is needed for successful minor surgery. An experienced assistant is also a great help. Sterile instruments and gloves, and aseptic technique are essential.

- **Basic minor surgery sets** Usually contain a scalpel; several sizes of blade (e.g. sizes 11 and 15); toothed forceps; needle holder; fine scissors; artery forceps; skin hook; curette
- **Additional equipment required** Skin preparation liquid (e.g. chlorhexidine); local anaesthetic (e.g. lidocaine 1%); suitable-sized needles and syringes; sterile towels; swabs; sterile specimen pots; suture materials and dressings for the wound. For joint injection, ensure that you have steroid and local anaesthetic drawn up and suitably sized needles available before starting

⚠ Always make sure you know how many blades, sutures, needles, and swabs you have, and ensure that you have accounted for and safely disposed of them at the end of the procedure.

Consent ➲ p. 48. Patient consent for the procedure must be sought and recorded in the notes. This involves giving enough information about the procedure and other possible treatment options to allow the patient to make an informed decision about whether to proceed; the patient and consenting doctor should then both sign the consent form and the form should be filed in the patient's medical records.

Histological examination All tissue removed by minor surgery should be sent for histological examination unless there are exceptional or acceptable reasons for not doing so.

Documentation Maintain full, legible, accurate records. Include:

- History of the complaint
- Examination findings
- Diagnosis
- Full details of the procedure undertaken—include dose, batch number, expiry date and quantities of drugs, size and number of sutures
- Follow-up arrangements

If the patient is not registered with the practice undertaking the minor surgery, then a complete record of the procedure must be sent to the patient's registered practice for inclusion in the GP notes.

Follow-up and outcome Should be recorded in the patient's notes.

Advise the patient:

- What to expect after the procedure
- Precautions to take after the procedure
- When to return for suture removal
- Signs that would indicate the need for reconsultation
- About the expected recovery/healing time

Arrange a follow-up appointment (e.g. with a practice nurse) for all but the most straightforward procedures. Ensure that histology results are checked and communicated to the patient.

Further information

Primary Care Dermatology Society Skin surgery guidelines. 🖱 www.pcds.org.uk/images/downloads/skin_surgery_guidelines.pdf

Basic techniques

⚠ Never attempt a procedure if you are unsure about it—know the boundaries of your experience and abilities.

Local anaesthesia

- 0.5–1% lidocaine is the most commonly used preparation due to its rapid onset of action and relatively short duration of effect
- Adrenaline/epinephrine (1:200,000) added to local anaesthetic ↓ bleeding and prolongs anaesthesia—but do not use adrenaline/epinephrine in areas supplied by end arteries (i.e. fingers, toes, penis, ear, nose), as vasoconstriction may cause ischaemia
- Safe maximum dose of lidocaine in adults is 20mL of 1% solution (less in the elderly/children)—overdose causes fits or cardiac arrhythmia

Administering local anaesthetic

- Pre-warn patients local anaesthetic stings before numbing and that they will still be able to feel pressure—but not pain. If pain is felt, more anaesthetic is needed
- Clean the skin, insert a small needle intradermally, and raise a small bleb before infusing more deeply; always pull back on the syringe plunger before injecting to check that you are not in a blood vessel—IV administration can result in fits and cardiac arrhythmia
- Anaesthetic must be infused all around the excision site. This may require several needle insertions—try to do this through an already numb area to ↓ discomfort for the patient; allow the anaesthetic to take effect (2–5min) before proceeding

Further information

Primary Care Dermatology Society Skin surgery—local anaesthetic. 🔗 www.pcds.org.uk/p/skin-surgery-local-anaesthetic

Suturing Various techniques for suturing and knot tying can be used (e.g. interrupted, continuous, mattress, subcuticular). Suture and needle types—➔ p. 136.

- Make a careful record of number of sutures and when they should be removed; usually, sutures need removal after 3–5d on the face, 7–14d on the back and legs, and 5–7d elsewhere
- Skin closure strips can be used instead of or in addition to sutures in some circumstances

Cautery Chemical (silver nitrate) or electrocautery are used alone, or in combination with other methods (e.g. curettage), to secure haemostasis or destroy tissue. Suitable conditions: nose bleeds, telangiectasia.

⚠ Do not use electrical cautery for patients with a cardiac pacemaker.

Implants Subcutaneous implants are prescribed for several conditions (e.g. prostate cancer). Most implants come pre-packaged with an insertion cannula and information leaflet—always read and follow the instructions if administering a new product and ensure position of implant and timing of administration is correct.

Contraceptive implants ➔ p. 739

Joint and soft tissue injection Steroids have potent anti-inflammatory effects and can dramatically improve certain musculoskeletal problems. Specific joint injections ➔ p. 138. Most joint injections are straightforward and can be undertaken in general practice. Hospital rheumatology departments usually have a joint injection clinic and often allow GPs to watch to gain experience.

General rules

- Always use an aseptic, no-touch technique
- Never inject into the substance of a tendon—this may cause rupture (in tenosynovitis steroid is injected into the tendon sheath)
- Injections should not require pressure on the syringe plunger—if so, the needle is not correctly located (tennis elbow is an exception)
- Undertake as few injections as possible to settle the problem—often 1 is sufficient; if no improvement after 2, reconsider the diagnosis
- Do no more than 1–2 injections/patient/appointment and no more than 3–4 injections/y in any single joint at intervals ≥3mo —more than this ↑ risk of systemic absorption and joint damage

> ⚠ *Do not inject if* Allergy to steroid or LA, local skin sepsis (e.g. cellulitis), any possibility of fracture or joint infection, prosthetic joint, severe joint destruction or unstable coagulopathy. If on warfarin, ensure INR <4.5 prior to the procedure; joint injections are safe for patients taking direct oral anticoagulants, e.g. rivaroxaban—avoid injecting during peak drug activity i.e. for 2–4h after drug administration

Preparation for the procedure

- Take a history, examine, and have a clear diagnosis before injecting
- Gather needles, syringes, a sterile container if needed (for sending aspirated fluid), steroid, local anaesthetic, skin preparation fluid (e.g. chlorhexidine or surgical spirit), cotton wool, and adhesive spot plaster
- Make sure the patient is comfortable and has given informed consent
- The joint should be rested for 2–5d after injection

Steroid preparations (↑ order of potency) hydrocortisone acetate, methylprednisolone acetate, triamcinolone acetonide.

Local anaesthetic (LA) (e.g. lidocaine 1%) can be injected first or mixed with the steroid for some injections—LA effect occurs immediately and lasts 2–4h. The patient may then experience some return of symptoms (pain) before the steroid takes effect—warn the patient.

Follow-up

- Some injections are painful at administration—this is normal for tennis elbow and plantar fasciitis
- Steroid injection may cause temporary flushing (mainly in ♀), menstrual irregularity, and worsening of blood sugar control in patients with DM
- Severe or increasing pain ~48h after injection may indicate sepsis—advise the patient to seek medical attention urgently if this occurs
- If steroid is injected close to the skin surface (as in tennis elbow), skin dimpling and pigment loss can occur—warn the patient

Further information

Silver T (2018) *Joint and Soft Tissue Injection: Injecting with Confidence* (6th edn). Boca Raton, FL: CRC Press. ISBN 9781138604179.

Removal of skin lesions

⚠ Rules for removing skin lesions

- Ensure that you have had formal training in all techniques required—learning by experience is much better than from a book. There are many courses available
- Only remove benign lesions (unless you have specific training to excise low-risk basal cell carcinomas)—refer suspicious lesions to a specialist for expert management
- Only remove lesions that you are confident you can cope with. Take special care with children, and lesions on the face or lip margin—the scar may be very noticeable
- Send all excised lesions for histology—place in formalin; label carefully

Surgical excision of skin lesions Gain written consent—ensure that you have warned the patient about the likely size of the scar and the possibility of keloid (especially if the lesion is on a risk area, e.g. upper back and chest). Work out the direction of the skin contour lines, clean and anaesthetize the area (➔ p. 134). An elliptical incision ~3× as long as it is wide is suitable for most lesions. Place the incision in the skin contour lines if possible (marking the incision line can be helpful). Cut through the skin at right angles to the surface with a smooth sweep of the blade. Use a skin hook to lift the skin from one end of the ellipse. Use the scalpel blade to remove the skin from the subcutaneous fat. Save the excised specimen for histology.

Close the wound by carefully opposing the edges (slightly everted) using non-absorbable sutures. Avoid tension in the sutures and knot securely. Large wounds may benefit from the use of deep absorbable sutures to reduce skin tension. Suture types—Box 6.1

Box 6.1 Suture types

Suture material

- **Absorbable** e.g. catgut, Dexon, Vicryl—used to stitch deep layers to help ↓ tension on the skin sutures
- **Non-absorbable** e.g. silk, Prolene, nylon—used for closure of skin wounds after minor surgery

Needle type Straight, curved, cutting, or round-bodied. Surgical site and personal preference dictate which to use—a cutting needle is usually used for skin.

Suture thickness (gauge) Indicated by a number (10/0 is fine and 2/0 thick). For skin closure: 6/0 or 5/0 is usually used for the face, 3/0 on legs and back, and 4/0 elsewhere.

Curettage Useful for seborrhoeic keratoses, pyogenic granuloma, or single viral warts. Not suitable for naevi. Use only if the diagnosis is certain—scrapings can be sent for histology but architecture of the lesion is lost. Clean the area, then numb with local anaesthetic Remove the lesion with gentle scooping movements using a curette spoon. Finally cauterize the base of the lesion.

Punch biopsy Allows histological examination of the skin for diagnostic purposes. Do not biopsy pigmented lesions in primary care. Choose biopsy site to include the skin changes of concern and minimize cosmetic impact. Clean and anaesthetize the area. Sterile, disposable punch biopsy tools 2–4mm diameter are suitable for use in primary care. Using aseptic technique, hold the tool perpendicular to the skin and gently rotate until a 'give' is felt as the tool enters the subcutaneous fat. Withdraw the punch and remove the specimen with forceps to send for histology. If needed, close the skin with skin closure strips or a suture.

Shave excision Suitable for removal of superficial benign skin lesions in primary care, e.g. prominent moles or skin tags. Clean and anaesthetize the area. Using aseptic technique, remove the lesion using a razor blade, scalpel, or shave excision instrument skimmed parallel to and on the surface of the skin. No skin closure is required. The excision site heals with a scab over several days. Send excised tissue for histology.

Cryotherapy Liquid nitrogen can be used to treat viral warts, and seborrhoeic and solar keratosis. Local arrangements for delivery of liquid nitrogen differ—often a clinic session to treat all suitable lesions at the same time is helpful. If diagnosis is uncertain excise the lesion, or take a biopsy prior to freezing. A cotton wool bud or nitrogen spray gun can be used to apply liquid nitrogen for approximately 10 seconds until a thin frozen halo appears at the base of the lesion. A blister forms <24h after treatment—the lesion then falls off with the blister. Repeat treatment may be needed after ≥4wk. Side effects include pain, failure to remove the lesion, skin hypopigmentation, and ulceration of lower leg lesions (especially in elderly patients).

Further information

Primary Care Dermatology Society Skin surgery series:

- Curettage 🖱 www.pcds.org.uk/p/skin-surgery-curettage
- Elliptical excision 🖱 www.pcds.org.uk/p/skin-surgery-elliptical-excision
- Epidermoid (sebaceous) cyst removal 🖱 www.pcds.org.uk/p/skin-surgery-epidermoid-sebaceous-cyst-removal
- Lipoma 🖱 www.pcds.org.uk/p/skin-surgery-lipoma
- Mattress suturing 🖱 www.pcds.org.uk/p/skin-surgery-mattress-suturing
- Punch biopsy 🖱 www.pcds.org.uk/p/skin-surgery-punch-biopsy
- Shave excision 🖱 www.pcds.org.uk/p/skin-surgery-shave-excision
- Subcutaneous suturing 🖱 www.pcds.org.uk/p/skin-surgery-subcutanous-suturing
- Subcuticular suturing 🖱 www.pcds.org.uk/p/skin-surgery-subcuticular-suturing
- Surface suturing 🖱 www.pcds.org.uk/p/skin-surgery-surface-suturing
- Suture knot 🖱 www.pcds.org.uk/p/skin-surgery-suture-knot

Commonly performed injections

The knee Joint effusions are common (e.g. trauma, ligament strains, OA, RA, gout). Aspirated fluid should be clear/slightly yellow and not purulent. Send any fluid aspirated for analysis. Aspiration of fluid can:

- Help make a diagnosis, e.g. gout
- Be a therapeutic procedure—draining a tense effusion can relieve pain
- Precede administration of steroids, e.g. RA flare

⚠ Any sign of infection within the joint prohibits steroid use.

Technique for aspiration and joint injection

- Lie the patient on couch with knee slightly bent—place a pillow under the knee as this relaxes the muscles
- Palpate the joint space under the lateral or medial edge of the patella and inject/aspirate just below the superior border of the patella with the needle horizontal—Figure 6.1
- Use a green (21-gauge) needle
- If aspirating and then injecting steroids, maintain the needle in position and swap the syringe
- Normal doses of steroid are triamcinolone acetonide 40mg or methylprednisolone acetate 40mg
- In pre-patellar bursitis, aspiration and injection of hydrocortisone 25mg into the bursa can help settle inflammation

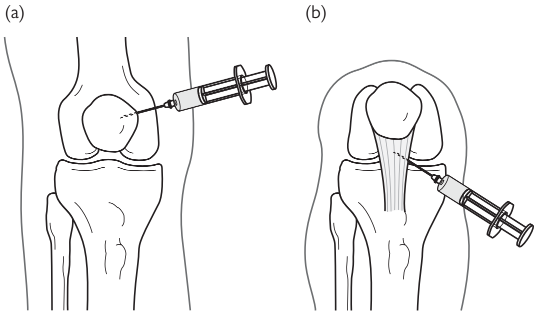

Figure 6.1 Knee joint injection or aspiration

Trochanteric bursitis Tender area overlying the greater trochanter of the hip. Steroid injection can alleviate pain.

Technique

- Lie the patient on one side with both hips and knees bent, so that the painful hip is uppermost; draw up triamcinolone acetonide 40mg and 5mL of 1% lidocaine
- Ask the patient to show you the most tender spot and mark it; then clean the skin and insert the needle (green, 21-gauge) perpendicular to the skin at the marked spot

- Insert as far as the needle allows or to bone, withdraw 3–5mm and inject 1mL; without taking the needle out, move the needle in a circle injecting 1mL in each direction—try to inject all the tender area

Plantar fasciitis Painful area in the middle of the heel pad. Steroid injection (e.g. triamcinolone acetonide 10–40mg) into the most tender spot can help. Injection hurts, so advise analgesia. Mixing lidocaine 1% with the steroid can help.

Technique Two methods are commonly used (Figure 6.2):
- Injection through the tough skin of the sole (more accurate), or
- Lateral approach (less painful)

Rest the foot for several days afterwards and use an in-shoe heel pad. Rupture of the plantar fascia is a rare complication.

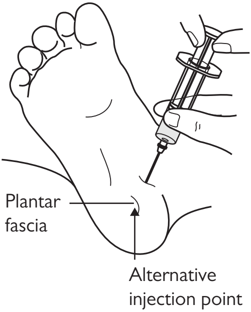

Figure 6.2 Injection of plantar fasciitis

Tenosynovitis Causes pain and stiffness in the line of the tendon and crepitus over the affected tendon. The most common site is the base of the thumb (de Quervain's tenosynovitis). Injecting steroid and LA (e.g. hydrocortisone 25mg + 1mL 1% lidocaine) into the space between tendon and sheath can help.

Technique
- Insert the needle along the line of the tendon just distal to the point of maximum tenderness
- Advance the needle proximally into the tendon (felt as a resistance) and then slowly withdraw until the resistance disappears. The tip of the needle is then in the tendon sheath
- It is now safe to inject—the tendon sheath may swell
- Advise the patient to rest the affected area for several days and avoid the precipitating activity

Carpal tunnel syndrome ➔ p. 460

Technique

- Sit the patient with hand resting on a firm surface, palm up. Palmaris longus tendon can be seen by wrist flexion against resistance
- Insert the needle at the distal skin crease, at 45° to the horizontal, pointing towards the fingers, just ulnar (little finger) side of the palmaris tendon (Figure 6.3). If palmaris longus is absent (10%), inject between flexor digitorum superficialis and flexor carpi radialis tendons
- Use a blue (23-gauge) needle. Advance it to half its length. If sudden pain in the fingers, you have hit the median nerve—withdraw the needle and reposition it
- Inject steroid, e.g. 10mg triamcinolone acetonide—if there is resistance the needle is not in the right place. Do not use LA as it causes finger numbness
- Rest the hand for several days afterwards

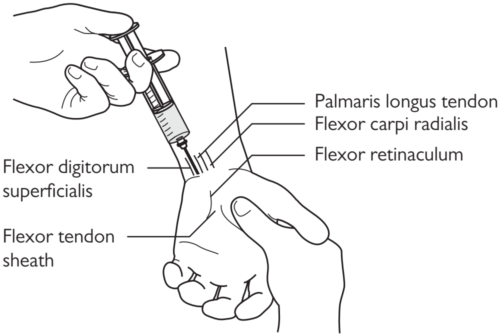

Figure 6.3 Injection of the carpal tunnel

Elbow

Technique for tennis/golfer's elbow injection

- Sit the patient with the elbow flexed to 90°
- Palpate the most tender spot and insert the needle into that spot
- Inject 0.1–0.2mL of steroid (e.g. hydrocortisone 25mg/mL). There will be resistance. Without taking the needle out, move the needle in a fan shape injecting small amounts of steroid—try to inject all the tender area. Warn about the possibility of skin dimpling or pigment loss
- Pain of injection may last 48h—warn the patient in advance, advise resting the arm and analgesia

💣 For tennis elbow, steroid injection has significantly better effects in the short term (~6wk) but poorer outcome long-term compared to physiotherapy[R].

Shoulder injection May help rotator cuff problems, impingement, frozen shoulder, subacromial bursitis, and RA. Use an anterior or posterior approach for shoulder joint injection and lateral approach for the subacromial space.

Technique: anterior approach

- Sit the patient with the arm relaxed at the side, slightly externally rotate
- Insert the needle (green, 21-gauge) horizontally into the gap between the head of humerus and the coracoid process, ensuring the needle is lateral to the coracoid process—Figure 6.4(a). Insert the needle for most of its length to reach the joint space
- Inject 1mL steroid, e.g. triamcinolone acetonide 40mg + 5mL 1% lidocaine
- There should be no/little resistance to injection—if there is, the needle is wrongly positioned

Technique: lateral approach to subacromial space Sit the patient with arm hanging down to the side. Palpate the posterolateral corner of the acromion. Insert the needle horizontally into the space under the acromion—Figure 6.4(b). Inject 40mg triamcinolone acetonide + 5mL 1% lidocaine.

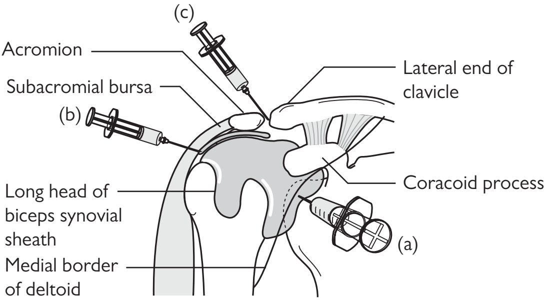

Figure 6.4 Shoulder joint injection

Technique: AC joint injection Can help the pain of OA.

- Palpate the joint space—insert the needle anteriorly or superiorly—if you go too far, you may enter the shoulder joint—Figure 6.4(c)
- Only 0.2–0.5mL of steroid can be injected as small joint space. Use a blue (23-gauge) needle and do not add LA

⚠ Warn patients that pain may worsen for up to 48h after injection before it improves.

Patient information

Arthritis Research UK Patient information: 'Local steroid injections'.
☎ 0800 5200 520 🖱 www.arthritisresearchuk.org

Chapter 7

Healthy living

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Prevention of travel-related illness *146*
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Management of alcohol misuse *160*
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Prevention and screening

In all disease, the goal is prevention.

Definitions

- **Primary prevention** Prevention of disease occurrence, e.g. childhood vaccinations; prophylactic mastectomy for women with BRCA1/2 mutations
- **Secondary prevention** Controlling disease in early form, e.g. cervical cancer and bowel cancer screening; AAA screening
- **Tertiary prevention** Prevention of complications once the disease is present, e.g. screening for AF in order to anticoagulate

Barriers to prevention

- **Patient** Blinkering ('*It'll never happen to me*'); rebellion ('*I know it's bad—but it's cool*'); poor motivation (path of least resistance)
- **Doctor** Time; money—health promotion takes time and personnel; motivation—health promotion is repetitive and boring
- **Society** Pressure from big business (e.g. cigarette advertising); other priorities; ethics (e.g. public uproar at threats not to offer cardiac surgery to smokers)

Screening The idea of screening is attractive—the ability to diagnose and treat a potentially serious condition at an early stage when it is still treatable. An ideal screening test must pick up all those who have the disease (have high sensitivity) and must exclude those who do not (high specificity). It must detect only those who have a disease (high positive predictive value) and should exclude only those who do not have the disease (high negative predictive value). See Table 7.1.

The Wilson–Jungner criteria All screening tests should meet the following criteria before they are introduced to the target population:

- The condition being screened for is an important health problem
- Natural history of the condition is well understood
- There is a detectable early stage
- Treatment at early stage is of more benefit than at late stage
- There is a suitable test to detect early-stage disease
- The test is acceptable to the target population
- Intervals for repeating the test have been determined
- Adequate health service provision has been made for the extra clinical workload resulting from screening
- Risks, both physical and psychological, are < benefits (Table 7.2)
- Costs are worthwhile in relation to benefits gained

UK screening programmes

- >40y health checks—➔ p. 214
- Diabetic retinopathy—➔ p. 328
- Childhood—➔ p. 826
- Neonatal bloodspot—➔ p. 830
- Neonatal hearing—➔ p. 836
- Screening for hip dysplasia—➔ p. 834
- Antenatal—➔ p. 782
- Chlamydia—➔ p. 716
- Cervical cancer—➔ p. 704
- Breast cancer—➔ p. 670
- Colon cancer—➔ p. 368
- Abdominal aortic aneurysm—➔ p. 254

Table 7.1 Screening test performance

	Disease present	Disease absent
Screening test positive	a	b
Screening test negative	c	d

- Sensitivity = $a/(a+c)$
- Specificity = $d/(b+d)$
- Positive predictive value = $a/(a+b)$
- Negative predictive value = $d/(c+d)$

Performance of screening tests See Table 7.1. For a screening programme to be effective and ↓ morbidity and mortality there must be:

- Adequate participation of the target population
- Few false-negative or false-positive results
- Screening intervals shorter than the time taken for the disease to develop to an untreatable stage
- Adequate follow-up of all abnormal results
- Effective treatment at the stage detected by screening

❗ There is no ideal screening test. Always explain:

- Purpose of screening
- Likelihood of positive/negative findings and possibility of false-positive/negative results
- Uncertainties and risks attached to the screening process
- Significant medical, social, or financial implications of screening for the particular condition or predisposition
- Follow-up plans, including availability of counselling/support services

Prevention of coronary heart disease ➔ p. 214

Table 7.2 Benefits and disadvantages of screening

Benefits	Disadvantages
• Improved prognosis for some cases detected by screening	• Longer morbidity in cases where prognosis is unaltered
• Less radical treatment for some early cases	• Overtreatment of questionable abnormalities
• Reassurance for those with negative test results	• False reassurance for those with false-negative results
• Increased information on natural history of disease and benefits of treatment at early stage	• Anxiety and sometimes morbidity for those with false-positive results
	• Unnecessary intervention for those with false-positive results
	• Hazard of screening test
	• Diversion of resources to the screening programme

Further information

Wilson JMG, Jungner G (1968) *Principles and Practice of Screening for Disease*. Public Health Paper No. 34. Geneva: World Health Organization.

Prevention of travel-related illness

UK residents going abroad The UK has reciprocal agreements with some other countries for the provision of urgently needed medical treatment at ↓ cost or free of charge. Countries and the services available are listed on the NHS website (🖱 www.nhs.uk). Only urgently needed treatment is provided on the same terms as for residents of that country. Travellers should always ensure that they have adequate travel insurance when going abroad.

Pre-travel assessment 8wk pre-departure where possible. *Check*:

- Age
- Purpose of travel
- Type of accommodation
- Current vaccination status
- General health
- Where and when intending to travel (including areas within a country and stopovers elsewhere)
- Previous experience of travel (including experience with antimalarials)

Health risks of travel

- Environmental hazards (e.g. changes in altitude/climate) Avoid rapid changes of altitude—take time to readjust; avoid sunburn. Advise ♀ taking combined hormonal contraception and trekking to altitudes of >4500m for >1wk to consider an alternative method of contraception
- Accidents Avoid potentially dangerous tasks under the influence of alcohol, e.g. swimming, driving. Avoid motorbikes—especially without helmets and protective clothing
- Illness abroad MI causes 61% of deaths related to international travel. Advise patients not to travel if unwell; to ensure adequate insurance including repatriation costs; to take enough supplies of regular medication when travelling to last the entire trip; and to take preventative steps to avoid infection
- Transport-related problems:
  - Fitness to fly ➔ p. 100
  - Jet lag
  - Motion sickness—advise to take OTC medication if afflicted
  - DVT—on flights >3h: advise to drink plenty of water, avoid alcohol, regularly get up and walk around, and consider prophylactic support stockings
- Psychological effects of travel

Vaccination 4% of deaths related to travel are due to infectious disease—advise patients to ensure they are fully vaccinated for the areas they are intending to visit. Information is available from Travax (🖱 www.travax.nhs.uk)—registration is needed.

Prevention of travellers' diarrhoea 50% of travellers experience some diarrhoea. Most cases last 4–5d. 1–2% last >1mo.

- Take care to eat and drink uncontaminated food and water
- Food should be freshly cooked and hot
- Avoid salads and cold meats/fish
- Eat fruit that can be peeled
- Stick to drinks made with boiling water or bottled drinks and water with an intact seal; avoid ice in drinks
- Use water purification tablets if necessary

Action If diarrhoea occurs when abroad, advise to use oral rehydration fluids (made up with fresh, boiled water), and only to take antidiarrhoeals if impossible to get to a toilet. Seek medical advice if blood in stool, fever, or not resolving in <72h (24h for the elderly or infants). ⚠ Warn not to use antidiarrhoeals if blood in stool, fever, or child <10y.

Prevention of malaria

- Awareness of risk High-risk areas are Central and South America; South East Asia; Pacific islands; sub-Saharan Africa—however brief the time there. Pregnant and asplenic patients are at particular risk
- Reduce the risk of mosquito bites Mosquitoes bite at night:
  - *Accommodation*—sleep in screened accommodation spraying screens with insecticide each evening and use a pyrethroid vaporizer. If screens are not available, use a permethrin-impregnated bed net (kits are available)
  - *Person*—in the evenings wear long-sleeved shirts and trousers; protect limbs with diethyltoluamide-containing repellent
  - *Chemoprophylaxis*—Table 7.3. Regimens vary with location and time of year. Information is available via Travax (🖱 www.travax.nhs.uk—registration needed)
- Awareness of residual risk Chemoprophylaxis is not 100% effective. Advise to seek medical advice if unwell for 6mo after return. ⚠ Malaria is a great mimic—have a high level of suspicion

Table 7.3 Chemoprophylactic antimalarial drugs

Drug	Dose	Start	Stop
Chloroquine	310mg/wk	1wk before entering malaria area	4wk after leaving malaria area
Mefloquine	250mg/wk	**2.5wk** before entering malaria area	4wk after leaving malaria area
Proguanil	200mg/d	1wk before entering malaria area	4wk after leaving malaria area
Proguanil + atovaquone	1 tablet/d	1–2d before entering malaria area	1wk after leaving malaria area
Doxycycline	100mg/d	1wk before entering malaria area	4wk after leaving malaria area

Prevention of HIV and hepatitis B and C

Advise:

- Avoid casual sexual contacts. If these occur use barrier methods of contraception (male and female condoms)
- Avoid shared needles (e.g. tattooing/ear piercing/drugs)
- Medical kits—if travelling to high-risk areas, take a clearly labelled medical kit containing sutures, syringes, and needles for emergency use
- Avoid blood transfusion. Most blood donations in the developing world are unscreened. Know your blood group. In an emergency, the Blood Care Foundation can arrange screened blood to be provided anywhere in the world (☎ 01403 262652; 🖱 www.bloodcare.org.uk)
- Vaccination for hepatitis B prior to travelling

Further information

Fit for Travel 🖱 www.fitfortravel.nhs.uk
Travax 🖱 www.travax.nhs.uk—registration needed.

Diet

The role of the GP and primary care team

- **Screening** Identification of obese patients and patients in need of dietary advice for other reasons
- **Assessment** Current diet, motivation, and barriers to change
- **Discussion and negotiation** Exploration of knowledge about diet; negotiation of goals
- **Goal setting** Provide information and 2–3 food-specific goals on each occasion—set a series of mini-targets that appear realistic and achievable; tailor them to existing diet and usual schedule
- **Monitoring progress**

Barriers to a good diet

- **Ignorance** Posters in surgeries, leaflets, screencasts on waiting room screens, and/or information on the practice website may help
- **Cultural differences** Modify information to be relevant; provide information in multiple languages as appropriate
- **Enjoyment** Perception of healthy diet as not enjoyable
- **Poverty** Fresh fruit/vegetables and lean meat/fish are expensive—some elements are cheap, e.g. potatoes, pasta, rice
- **Lifestyle** Convenience foods contain a lot of salt, sugar, and fat
- **Peer pressure** Children are under pressure to eat sweets, crisps, etc.
- **Habits of a lifetime** We like the foods we have grown up with
- **Confusion about what is good** Packaging may be misleading, e.g. breakfast cereals claiming health messages but containing high sugar
- **Mixed messages** One minute the press says something is good for you, the next it causes some horrible disease and should be avoided
- **Fatalism/apathy**

The ideal diet See Figure 7.1, ➔ p. 150, Adjust composition/portion size of each meal to maintain a healthy weight. Include a variety of foods:

- Use starchy foods (e.g. bread, rice, pasta, potatoes) as the main energy source
- Eat plenty of fruit and vegetables (>5 portions of fruit/vegetables daily); do not overcook vegetables—steaming is preferable to boiling, and keep delay between cutting and eating fruit/vegetables to a minimum
- Eat plenty of fibre—good sources are high-fibre breakfast cereals, beans, pulses, wholemeal bread, potatoes (with skins), pasta, rice, oats, fruit/vegetables
- Eat fish at least 2×/wk including 1 portion (maximum 2 portions if pregnant) of oily fish (e.g. mackerel, herring, pilchards, salmon)
- Cut back on cooked red or processed meat; consider substituting meat with vegetable protein (e.g. pulses, soya)
- Choose lean meat—remove excess fat/poultry skin and pour off fat after cooking; avoid fatty meat products (e.g. sausages, salami, meat pies); boil, steam, or bake foods in preference to frying; when cooking with fat use unsaturated oil (e.g. olive, sunflower oil) and use cornflour rather than butter and flour to make sauces
- Use skimmed milk and low-fat yoghurts/spreads/cheese (e.g. Edam or cottage cheese)
- Avoid adding salt to foods—aim for <6g of salt/d. Avoid processed foods, crisps, and salted nuts

- Avoid adding sugar and cut down on sweets, biscuits, and desserts
- Drink at least 4–6 pints (2–3L) of fluid daily—preferably not tea, coffee, or alcohol. Drinking a large glass of water with meals and instead of snacks can reduce the urge to overeat
- Avoid excessive alcohol intake—<14U/wk for ♂ and ♀—➲ p. 158
- Obesity ➲ p. 152

Unintentional weight loss Non-specific symptom. Treat the cause. *Consider*:

- **GI causes**—malabsorption, malnutrition, dieting
- **Chronic disease**—hyperthyroidism, DM, heart failure, renal disease, severe COPD, degenerative neurological/muscle disease, chronic infection (e.g. TB, HIV) or infestation
- **Malignancy**
- **Psychiatric causes**—depression, anxiety, dementia, anorexia

Malnutrition 50% of ♀ and 25% of ♂ aged >85y are unable to cook a meal alone. Malnutrition is common amongst the elderly.

Poor nutritional status Slows rate of wound healing, ↑ risk of infection, ↓ muscle strength, is detrimental to mental well-being, and ↓ the ability of elderly people to remain independent.

Risk factors

- Low income
- Living alone
- Dementia
- Recent bereavement
- Mental health problems e.g. depression
- Gastric surgery
- Malabsorption
- ↑ metabolism e.g. thyrotoxicosis
- Difficulty eating and/or swallowing, e.g. stroke, MND
- Presence of chronic disease, e.g. Crohn's disease, UC, IBS, cancer, COPD, CCF

Management

- **General advice**—encourage to eat more and ↑ consumption of fruit and vegetables; consider using nutritional supplements, e.g. vitamin D supplements for the housebound and institutionalized
- **Inability to prepare meals/shop**—consider referral to social services, meals on wheels, community dietician; community day centre; local voluntary support organization
- **Difficulty with utensils**—aids or adapted equipment may help, e.g. special cutlery, non-slip mats—consider OT referral
- **Nausea**—consider antiemetics
- **Swallowing difficulty**—investigate the cause. If none is found or you are unable to resolve the problem, consider pureed food and/or thickened fluids—take dietician advice

Further information

British Nutrition Foundation 🖱 www.nutrition.org.uk

Malnutrition Universal Screening Tool (MUST) www.bapen.org.uk/screening-for-malnutrition/must/introducing-must

NICE (2006, updated 2017) Nutrition support for adults. 🖱 www.nice.org.uk/Guidance/cg32

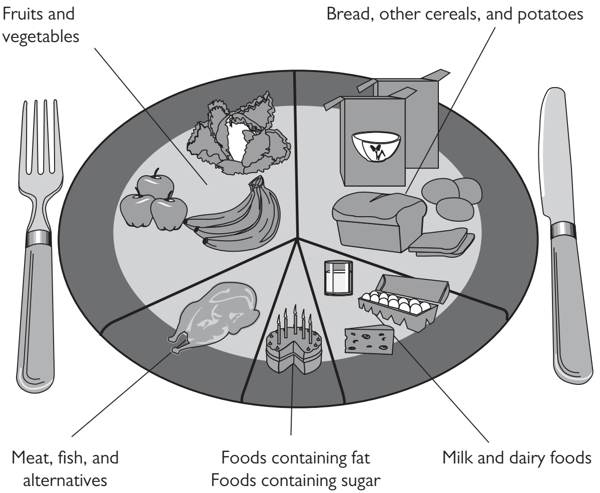

Fruits and vegetables
Bread, other cereals, and potatoes
Meat, fish, and alternatives
Foods containing fat
Foods containing sugar
Milk and dairy foods

Figure 7.1 The plate model. Developed to communicate current recommendations for healthy eating. It shows rough proportions of the various food groups that should make up each meal

What is a portion of fruit or vegetables? 1 portion of fruit or vegetables is roughly equivalent to:

- 1 normal portion (2 tablespoons) of any vegetable
- 1 dessert bowl of salad
- 1 large fruit, e.g. apple, banana, orange, pear, peach, large tomato, or a large slice of pineapple or melon
- 2 smaller fruits, e.g. satsumas, plums, kiwi fruits, apricots
- 1 cup of small fruits, e.g. strawberries, raspberries, blackcurrants, cherries, grapes
- 1 tablespoon of dried fruit
- 2 large tablespoons of fruit salad or stewed/canned fruit in natural juice
- 1 glass (150mL) of fresh fruit juice

Tips for avoiding snacking Discourage uncontrolled snacking of junk food between meals. Suggest patients ask themselves the following questions when they feel like eating between meals:

Am I hungry? If unsure, wait 20min, and then ask the same question again.

When was the last time I ate? If <3h ago, it may not be real hunger.

Could a small snack tide me over until the next meal? Have ready-to-eat fruits or vegetables on hand for this.

Height in metres

Weight in kilograms	1.36	1.40	1.44	1.48	1.52	1.56	1.60	1.64	1.68	1.72	1.76	1.80	1.84	1.88	1.92	1.96	2.00
125	68	64	60	57	54	51	49	46	44	42	40	39	37	35	34	33	31
123	67	63	59	56	53	51	48	46	44	42	40	38	36	35	33	32	31
121	65	62	58	55	52	50	47	45	43	41	39	37	36	34	33	31	30
119	64	61	57	54	52	49	46	44	42	40	38	35	35	34	32	31	30
117	63	60	56	53	51	48	46	44	41	40	38	36	35	33	32	30	29
115	62	59	55	53	50	47	45	43	41	39	37	35	34	33	31	30	29
113	61	58	54	52	49	46	44	42	40	38	36	35	33	32	31	29	28
111	60	57	51	51	48	46	43	41	39	38	36	34	33	31	30	29	28
109	59	56	50	50	47	45	43	41	37	37	35	34	32	31	30	28	27
107	58	55	52	49	46	44	42	40	38	36	35	33	32	30	29	28	27
105	57	54	51	48	45	43	41	39	37	35	34	32	31	30	28	27	26
103	56	53	50	47	45	42	40	38	36	35	33	32	30	29	28	27	26
101	55	52	49	46	44	42	39	38	36	34	33	31	30	29	27	26	25
99	54	51	48	45	43	41	39	37	35	33	32	31	29	28	27	26	25
97	52	49	47	44	42	40	38	36	34	33	31	30	28	27	26	25	24
95	51	48	46	43	41	39	37	35	34	32	31	29	28	27	26	25	24
93	50	47	45	42	40	38	36	34	33	31	30	29	27	26	25	24	23
91	49	46	44	42	39	37	36	34	32	31	29	28	27	26	25	24	23
89	48	45	43	41	39	37	35	33	32	30	29	27	26	25	24	23	22
87	47	44	42	40	38	36	34	32	21	29	28	27	26	25	24	23	22
85	46	43	41	39	37	35	33	32	30	29	27	26	25	24	23	22	21
83	45	42	40	38	36	34	32	31	29	28	27	26	25	23	23	22	21
81	44	41	39	37	35	33	32	30	29	27	26	25	24	23	22	21	20
79	43	40	38	36	34	32	31	29	28	27	26	24	23	22	21	21	20
77	42	39	37	35	33	32	30	29	27	26	25	24	23	22	21	20	19
75	41	38	36	34	32	31	29	28	27	25	24	23	22	21	20	20	19
73	39	37	35	33	32	30	29	27	26	25	23	23	22	21	20	19	18
71	38	36	34	32	31	29	28	26	25	24	23	22	21	20	19	18	18
69	37	35	33	32	30	28	27	26	24	23	22	21	20	20	19	18	17
67	36	34	32	31	29	28	26	25	24	23	22	21	20	19	18	17	17
65	35	33	31	30	28	27	25	24	23	22	21	19	19	18	18	17	16
63	34	32	30	29	27	26	25	23	22	21	20	19	19	18	17	16	16
61	33	31	29	28	26	25	24	23	22	21	20	19	18	17	17	16	15
59	32	30	28	27	26	24	23	22	21	20	19	18	17	17	16	15	15
57	31	26	27	26	25	23	22	21	20	19	18	18	17	16	15	15	14
55	30	30	27	25	24	23	21	20	19	19	18	17	16	16	15	14	14
53	29	29	26	24	23	22	21	20	19	18	17	16	16	15	14	14	13
51	28	26	25	23	22	21	20	19	18	17	16	16	15	14	14	13	13
49	26	25	24	22	21	20	19	18	17	17	16	15	14	14	13	13	12
47	25	24	23	21	20	19	18	17	17	16	15	15	14	13	13	12	12
45	24	23	22	21	19	18	18	17	16	15	15	14	13	13	12	12	11
43	23	22	21	20	19	18	17	16	15	15	14	13	13	12	12	11	11

▨ BMI <18.5—underweight

▨ BMI 18.5–24.9—acceptable weight

▨ BMI 25–29.9—overweight

▨ BMI 30–39.9—obese

■ BMI ≥40—morbid obesity

Figure 7.2 Body mass index (BMI) ready reckoner for adults

For children Body mass index child reference tables should be used (available from www.health forallchildren.co.uk).

Definitions:

- Overweight—weight ≥91st centile
- Obese—weight ≥98th centile

Obesity

Obesity is one of the most important preventable diseases in the UK (see Box 7.1). The best measure of obesity is body mass index (BMI).

Classification BMI (weight in kg ÷ (height in m)^2) (Figure 7.2, ➔ p. 151):

- <18.5kg/m^2—underweight
- 18.5–24.9kg/m^2—healthy weight
- 25–29.9kg/m^2—overweight
- 30–34.9kg/m^2—obesity I
- 35–39.9kg/m^2—obesity II
- >40kg/m^2—obesity III or 'morbid obesity'

Waist circumference See Table 7.4. Alternative measure of body fat correlated with CVD risk, DM, hyperlipidaemia, and ↑ BP. Measured halfway between the superior iliac crest and the rib cage. Use in addition to BMI to aid assessment of health risks.

Table 7.4 Waist circumference with ↑ risk (RR ≥3) of CHD/DM

Waist circumference	White Caucasians	Asians
Male	≥102cm (40 inches)	≥90cm (36 inches)
Female	≥88cm (35 inches)	≥80cm (32 inches)

❗ For every 1cm ↑ in waist circumference, the RR of a CVD event ↑ by ~2%.

Causes of obesity

- Socioeconomic factors
- Physical inactivity
- Smoking cessation—mean weight ↑ 3–4kg
- Polygenic genetic predisposition (~1 in 3 obese people)—more prone to obesity again after successful dieting
- Childbirth—especially if not breastfeeding
- Drugs—steroids, antipsychotics (e.g. olanzapine), contraceptives (especially depo-injections), sulfonylureas, insulin
- Endocrine causes (rare) e.g. hypothyroidism, Cushing's syndrome, PCOS—only investigate if other symptoms/signs of endocrine disease
- Ongoing binge eating disorder (➔ p. 992)

Prevention Begins in childhood with healthy patterns of exercise/diet.

Management When the body's intake > output over a period of time, obesity results. Management aims to reverse this trend on a long-term basis through healthy diet, adjustment of calorie intake, physical exercise, and psychological support.

Initial assessment Assess willingness to change, eating behaviour and diet, physical activity, psychological distress, and social and family factors affecting diet. Check a baseline BMI and waist circumference. Check BP, blood glucose, and fasting lipid profile.

General advice Whether willing to change or not, provide advice on risks of obesity, and benefits of healthy eating (➔ p. 148) and physical exercise (➔ p. 154). Tailor your advice to the individual. If unwilling to change, reinforce this information at each subsequent encounter with the patient.

Diet Advise a weight loss diet for any patient who is overweight/obese and willing to change:

- **Low-calorie diet**—obese people do lose weight if they ↓ their energy intake. Aim for weight ↓ of 1–2lb (0.5–1kg)/wk using a ↓ in calorie

Box 7.1 Health risks of obesity

Greatly increased risk (RR >3)
- Mortality (BMI >30kg/m^2)
- Type 2 DM (BMI of 35kg/m^2 confers a 92× ↑ risk of DM)
- Gall bladder disease
- Dyslipidaemia
- Insulin resistance
- Breathlessness
- Sleep apnoea

Moderately increased risk (RR 2–3)
- CHD (5–6% of deaths are due to obesity)
- ↑ BP
- OA (knees)
- Hyperuricaemia/gout

Slightly increased risk (RR 1–2)
- Cancer (breast in post-menopausal women, endometrial, oesophageal, colon)—14–20% of cancer deaths are due to obesity
- Reproductive hormone abnormalities
- PCOS
- Impaired fertility
- Low back pain
- Stress incontinence
- Anaesthetic and postoperative risk
- Fetal defects associated with maternal obesity
- Suicide
- School/workplace prejudice

intake of ~600kcal/d, and a target BMI of 25 kg/m^2, in steps of 5–10% of original weight. There is no health benefit of weight ↓ below this. If simple diet sheets are not effective, refer to a dietician

- **Very low-calorie diets** (<1000kcal/d)—have only limited place in management—use for a maximum of 12wk for obese patients when weight loss has plateaued

Group and behavioural therapy Group activities, e.g. Weight Watchers, have higher success rates in producing/maintaining weight ↓. Behavioural therapy together with low-calorie diets is also effective.

Drug therapy Orlistat (120mg tds with food) is the only drug licensed for treatment of obesity in the UK. It acts by ↓ fat absorption. Consider a 3mo trial if supervised diet/exercise has failed and

- BMI ≥30kg/m^2 or
- BMI ≥27kg/m^2 + co-morbidity (e.g. DM, ↑ BP)

Continue treatment >3mo only if weight ↓ is ≥5% of initial body weight.

Surgery Consider if BMI >40kg/m^2, or BMI 35–39.9 kg/m^2 and suffering from a condition that could be improved by weight loss, and non-surgical measures have failed. Adjustable gastric banding is the most common procedure. *Complications*: band slippage/damage; gastric erosion; pouch dilatation; infection; malabsorption.

Maintenance of weight loss Once a patient has lost weight, continue to monitor diet. Ongoing follow-up helps to sustain weight loss. Weight fluctuation (yo-yo dieting) may be harmful.

Further information

National Obesity Forum 🖱 www.nationalobesityforum.org.uk
SIGN (2010) Management of obesity. 🖱 www.sign.ac.uk/sign-115-management-of-obesity.html
NICE (2014) Obesity: identification, assessment and management. 🖱 www.nice.org.uk/guidance/cg189

Exercise

In the UK, 60% of adults are not active enough to benefit their health.

Recommended amounts of activity

- Adults: ≥30min/d moderate intensity exercise on ≥5d/wk
- Children: ≥1h/d moderate intensity exercise every day

Assessing levels of physical activity See Figure 7.3. Use a validated tool to assess levels of physical activity, e.g. General Practitioner Physical Activity Questionnaire (GPPAQ).

Health benefits of exercise Regular physical activity:

↓ risk of

- DM—through ↑ insulin sensitivity
- Obesity—➔ p. 152
- Cardiovascular disease—physically inactive people have ~2× ↑ risk of CHD and ~3× ↑ risk of stroke
- Osteoporosis—exercise ↓ risk of hip fractures by ½
- Cancer— exercise ↓ risk of colon cancer by ~40%. There is also evidence of a link between exercise and ↓ risk of breast and prostate cancers

Is a useful treatment for

- ↑ BP—can delay onset of hypertension, and result in 10mmHg drop of systolic and diastolic BP in people with established hypertension
- Hypercholesterolaemia— exercise results in ↑ high-density lipoprotein (HDL), and ↓ low-density lipoprotein (LDL)
- Cardiac rehabilitation (➔ p. 230) and COPD (➔ p. 286)
- DM—exercise improves insulin sensitivity and favourably affects other risk factors for DM, including obesity, HDL/LDL ratio, and ↑ BP
- Arthritis and back pain—exercise maintains function
- Mental illness—exercise ↓ intensity of depression and ↓ anxiety

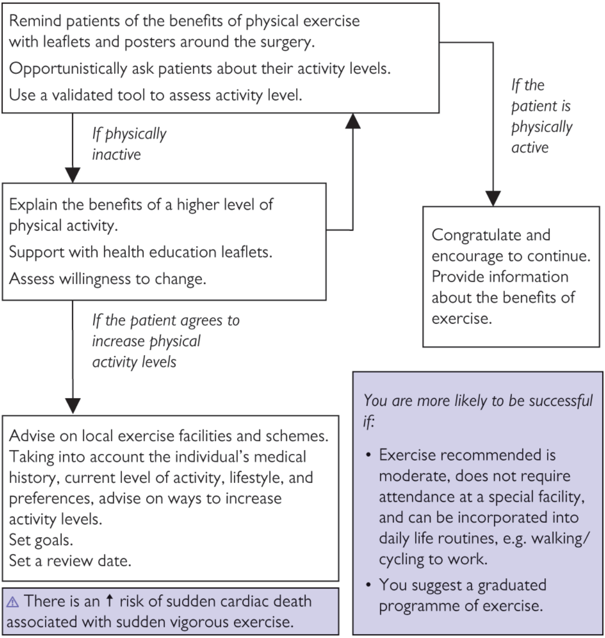
Benefits for the elderly

- Maintains functional capacity
- ↓ risk of falls and hip fracture
- ↓ levels of disability
- Improves quality of sleep

	Occupation			
Physical exercise and/or cycling (h/wk)	**Sedentary**	**Standing**	**Physical**	**Heavy manual**
0	Inactive	Moderately inactive	Moderately active	Active
Some but <1	Moderately inactive	Moderately active	Active	Active
1–2.9	Moderately active	Active	Active	Active
≥3	Active	Active	Active	Active

Figure 7.3 Physical activity index (PAI) derived from the GPPAQ

Reproduced from Department of Health and Social Care. *The General Practice Physical Activity Questionnaire (GPPAQ)*. © Crown copyright. Available at https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/192453/GPPAQ_-_guidance.pdf. Contains public sector information licensed under the Open Government Licence v3.0.



Remind patients of the benefits of physical exercise with leaflets and posters around the surgery.

Opportunistically ask patients about their activity levels.

Use a validated tool to assess activity level.

If physically inactive

Explain the benefits of a higher level of physical activity.

Support with health education leaflets.

Assess willingness to change.

If the patient agrees to increase physical activity levels

Advise on local exercise facilities and schemes. Taking into account the individual's medical history, current level of activity, lifestyle, and preferences, advise on ways to increase activity levels.

Set goals.

Set a review date.

⚠ There is an ↑ risk of sudden cardiac death associated with sudden vigorous exercise.

If the patient is physically active

Congratulate and encourage to continue. Provide information about the benefits of exercise.

You are more likely to be successful if:

- Exercise recommended is moderate, does not require attendance at a special facility, and can be incorporated into daily life routines, e.g. walking/cycling to work.
- You suggest a graduated programme of exercise.

Figure 7.4 Management plan for increasing activity levels

Effective interventions

- **Healthcare counselling** Is as effective as more structured exercise sessions (see Figure 7.4). Specialist rehabilitation schemes are available for patients with specific conditions (e.g. post-MI, COPD); exercise schemes operate in some areas, offering low cost, supervised exercise for patients who might otherwise find it unacceptable to visit a gym and are accessed via GP 'prescription'; many sports facilities offer special sessions for pregnant women, the over-50s, and people with disability
- **Workplace interventions** To ↑ rates of walking to work are effective
- **Schools** Appropriately designed and delivered physical education curricula can enhance physical activity levels. A whole-school approach to physical activity promotion is effective
- **Transport** Well-designed interventions ↑ walking/cycling to work
- **Communities** Community-wide approaches ↑ activity

Further information

DH (2011) UK physical activity guidelines 🖱 www.gov.uk/government/publications/uk-physical-activity-guidelines

NICE (2013) Physical activity: brief advice for adults in primary care. 🖱 www.nice.org.uk/guidance/ph44

Smoking

Facts and figures In England, 21% of adults (♂ 21%; ♀ 20%) smoke. Prevalence is highest amongst those aged 20–24y (32%) and lowest aged >60y (12%). 6% of school children aged 11–15y are regular smokers (♀ 10%; ♂ 8%). Surveys of smokers show 73% want to stop and 30% intend to give up in <1y—but only ~2%/y successfully give up permanently.

Risks of smoking Smoking is the greatest single cause of illness and premature death in the UK. Half of all regular smokers will die as a result of smoking—106,000 people/y. Smoking is associated with ↑ risk of:

- Cancers ~29% of all cancer deaths. Common cancers include: lung (>90% are smokers); lip; mouth; stomach; colon; bladder
- Cardiovascular disease CHD, CVA, peripheral vascular disease
- Chronic lung disease COPD, recurrent chest infection, exacerbation of asthma (29% of respiratory deaths result from smoking)
- Problems in pregnancy PET, IUGR, preterm delivery, neonatal and late fetal death
- DM
- Osteoporosis
- Thrombosis
- Dyspepsia ± gastric ulcer

Passive smoking is associated with

- ↑ risk of coronary heart disease and lung cancer (↑ by 25%)
- ↑ risk of cot death, bronchitis, and otitis media in children

Helping people to stop smoking Advice from a GP results in 2% of smokers stopping—5% if advice is repeated. See Figure 7.5.

Aids to smoking cessation

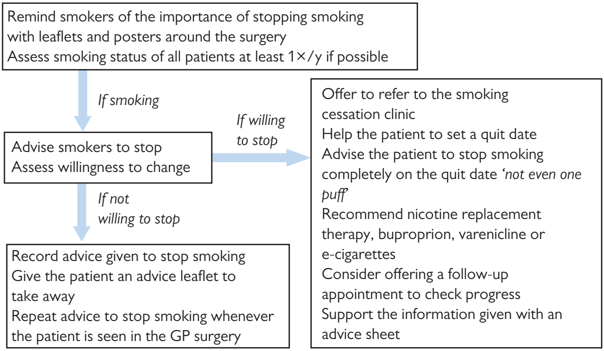
Nicotine replacement therapy (NRT) ↑ the chance of stopping ~1½×. All preparations are equally effective. Start with higher doses for patients highly dependent. Continue treatment for 3mo, tailing off dose gradually over 2wk before stopping (except gum which can be stopped abruptly). Contraindicated immediately post-MI, stroke, or TIA, and for patients with arrhythmia.

Bupropion Smokers (>18y) start taking the tablets 1–2wk before intended quit day (150mg od for 3d, then 150mg bd for 7–9wk). ↑ cessation rate >2×. *Contraindications*: epilepsy or ↑ risk of seizures, eating disorder, bipolar disorder.

Varenicline Smokers (>18y) start taking the tablets 1wk before intended quit day (0.5mg od for 3d, 0.5mg bd for 4d, then 1mg bd for 11wk). ↓ dose to 1mg od if renal impairment/elderly. ↑ cessation rate >2×. If the patient has stopped smoking after 12wk, consider prescribing a further 12wk treatment to ↓ chance of relapse. *Contraindications*: caution in psychiatric illness.

e-cigarettes Heat a liquid (usually comprising propylene glycol and glycerol ± flavours) into an aerosol for inhalation. Vary in nicotine content from none to >20mg/mL. Although good-quality evidence is currently lacking, the general consensus is that e-cigarettes do ↑ smoking cessation rates—both through nicotine replacement, and by addressing sensory/behavioural aspects of smoking addiction. e-cigarettes are not licensed as medicines currently and are not available on NHS prescription. Long-term effects of 'vaping' are as yet unclear.

Alternative therapies Hypnotherapy may be helpful in some cases.



Remind smokers of the importance of stopping smoking with leaflets and posters around the surgery
Assess smoking status of all patients at least 1×/y if possible

Advise smokers to stop
Assess willingness to change

Offer to refer to the smoking cessation clinic
Help the patient to set a quit date
Advise the patient to stop smoking completely on the quit date *'not even one puff'*
Recommend nicotine replacement therapy, buproprion, varenicline or e-cigarettes
Consider offering a follow-up appointment to check progress
Support the information given with an advice sheet

Record advice given to stop smoking
Give the patient an advice leaflet to take away
Repeat advice to stop smoking whenever the patient is seen in the GP surgery

Nicotine withdrawal symptoms:
Urges to smoke (70%)
Increased appetite (70%—mean 3–4kg weight ↑)
Depression (60%)
Restlessness (60%)
Poor concentration (60%)
Irritability/aggression (50%)
Night-time wakening (25%)
Light-headedness (usually just the first few days after quitting) (10%)

Figure 7.5 Suggested management plan for smoking cessation

Support In many areas, 'stop smoking' services are provided by PCOs. These programmes consist of a combination of group education, counselling ± individual support in combination with nicotine replacement, bupropion, varenicline, or e-cigarettes.

Prescribing smoking cessation medication Prescribe only for smokers who commit to a target stop date. Initially, prescribe only enough to last 2wk after the target stop date, i.e. 2wk nicotine replacement therapy, 3–4wk bupropion, or 3wk varenicline. Only offer a second prescription if the smoker demonstrates continuing commitment to stop smoking. ❗ If unsuccessful, the NHS will not fund another attempt for ≥6mo.

Smokeless tobacco Misri India tobacco, qimam, naswar, gul, khaini, gutkha, zarda, mawa, Manipuri, or betel quid with tobacco. Particularly used in South Asian communities. Carries risk of nicotine addiction, CVD, dental disease, and mouth/throat cancer. Provide brief advice to stop; consider NRT.

Further information

Hartmann-Boyce J, et al. (2018) Electronic cigarettes for smoking cessation. *BMJ* **360**:j5543.
NICE (2012) Smokeless tobacco cessation: South Asian communities. 🖱 www.nice.org.uk/guidance/ph39
NICE (2018) Stop smoking interventions and services. 🖱 www.nice.org.uk/guidance/ng92

Useful contacts

Action on smoking and health (ASH) 🖱 www.ash.org.uk
NHS Smokefree 🖱 www.nhs.uk/smokefree
Quit ☎ 0800 00 22 00 🖱 www.quit.org.uk

Alcohol

> *'An alcoholic is someone you don't like who drinks as much as you do'*
>
> Dylan Thomas (1914–1953)

Alcohol misuse is a major public health and social concern. Alcohol-related problems cost the NHS ~£1.7 billion/y. Most harm is caused by non-dependent drinkers. Screening (Figure 7.6) and brief interventions in primary care can identify drinkers in this group and ↓ consumption and harm.

What is a unit of alcohol? 1 unit = 10mL (or 8g) of pure alcohol. It is the amount of alcohol that an adult can process in ~1h—though speed of elimination does vary. It can be calculated. The 'alcohol by volume' (ABV) is stated on the packaging of all alcoholic drinks sold in the UK. Calculating units from ABV: number of units = ABV × volume (mL) ÷ 1000. As a rough guide, 1 unit ≈ ½ pint of beer, a small glass of wine, or a single shot of spirit.

Recommended limits ≤14U/wk for ♂ and ♀.

Prevalence of alcohol misuse

- Hazardous/harmful drinking—excess drinking causing potential or actual harm but without dependence—affects 32% ♂; 15% ♀
- Binge drinking (>8U for ♂ or >6U for ♀ in 1d)—affects 21% ♂; 9% ♀
- Alcohol dependence—affects 6% ♂; 2% ♀

Alcohol and health Moderate consumption (1–3U/d) may ↓ risk of non-haemorrhagic stroke, angina, and MI—but overall risks >> benefits. Risk depends on other factors too (e.g. smoking, heart disease). Potential harms:

Death 15,000–22,000 deaths/y in the UK are associated with alcohol misuse—most related to stroke, cancer, liver disease, accidental injury/suicide.

Physical health

- Obesity (high calorie content)
- Fatty liver
- Hepatitis
- Cirrhosis
- Liver cancer
- Oesophageal varices ± haemorrhage
- Gastritis
- Pancreatitis
- ↑ BP
- CVA
- DM
- Cancer of the mouth, larynx, and oesophagus
- Breast cancer
- Haemopoietic toxicity (↑ MCV)
- Nutritional deficiencies
- Neuropathy
- Myopathy
- Cardiomyopathy
- Poor sleep
- Tiredness
- Brain damage
- Sexual dysfunction
- Infertility
- Fetal damage
- Back pain
- Interactions with prescribed drugs
- Injuries due to alcohol-related activity (e.g. fights)

Mental health Anxiety, depression, and/or suicidal ideas; dementia and/or Wernicke's encephalopathy ± Korsakoff's syndrome (➔ p. 553).

Social harms of alcohol

- Marriage breakdown
- Loss of work
- Poverty
- Absence from work
- Social isolation
- Loss of shelter/home

Questions

1) How often do you have a drink containing alcohol?

Scoring:	*0*	Never	*3*	2–3×/wk
	1	1×/mo	*4*	≥4×/wk
	2	2–4×/mo		

2) How many drinks containing alcohol do you have on a typical day when you are drinking?

Scoring:	*0*	1 or 2 drinks	*3*	7–9 drinks
	1	3 or 4 drinks	*4*	≥10 drinks
	2	5 or 6 drinks		

3) How often do you have 6 or more drinks on one occasion?

4) How often during the last year have you found that you were not able to stop drinking once you started?

5) How often during the last year have you failed to do what was normally expected of you because of drinking?

6) How often during the last year have you needed a first drink in the morning to get yourself going after a heavy drinking session?

7) How often during the last year have you had a feeling of guilt or remorse after drinking?

8) How often during the last year have you been unable to remember what happened the night before because of your drinking?

Scoring:	*0*	Never	*3*	Weekly
	1	<1×/mo	*4*	Daily or almost daily
	2	Monthly		

9) Have you or someone else been injured because of your drinking?

10) Has a relative, friend, doctor or other health care worker been concerned about your drinking or suggested that you cut it down?

Scoring:	*0*	No
	2	Yes – but not in the last year
	4	Yes – in the last year

Questions assessing hazardous alcohol use	Questions assessing dependence symptoms	Questions assessing harmful alcohol use

Action*

Audit score 0–7	Alcohol education
Audit score 8–15	Alcohol education + simple advice
Audit score 16–19	Simple advice + brief counselling + continued monitoring
Audit score 20–40	Referral to specialist alcohol services for evaluation and treatment

* Provide the next highest level of intervention to patients who score ≥2 on Questions 4, 5 and 6, or 4 on Questions 9 or 10.

Figure 7.6 The alcohol use disorders identification test (AUDIT)

Reproduced with permission from Babor, T.F. *et al. The Alcohol Use Disorders Identification Test: Guidelines for Use in Primary Care*, second edition. Geneva, Switzerland: World Health Organization. Copyright © World Health Organization 2001. https://www.who.int/substance_abuse/publications/audit/en/

Management of alcohol misuse

Assessing for alcohol misuse

Screening Ask patients directly about their alcohol use. Use standardized questionnaires to identify patients with harmful and hazardous patterns of alcohol consumption, e.g. AUDIT (Figure 7.6, ➔ p. 159).

Suspicious signs/symptoms ↑ or uncontrolled BP; obesity; recurrent injuries/accidents; non-specific GI complaints; back pain; poor sleep; tiredness.

Risk factors

- Previous personal history of alcohol misuse
- Family history
- Poor social support
- Work absenteeism
- Emotional and/or family problems
- Financial and legal problems
- Drug problems
- Alcohol associated with work, e.g. publican

Examination Smell of alcohol, tremor, sweating, slurring of speech, ↑ BP, signs of liver damage.

Investigations FBC (↑ MCV); LFTs (↑ GGT in ~25% of heavy drinkers; ↑ AST; ↑ bilirubin). USS—fatty liver/cirrhosis. Often incidental findings.

Alcohol and driving Advise patients who persistently misuse alcohol that they must stop driving and notify the DVLA. Licence is revoked until ≥6mo of abstinence/controlled drinking (≥1y if Group 2 licence) and normalization of blood parameters.

Alcohol management strategies (Figure 7.7) For patients drinking within acceptable limits, reaffirm the limits. If misusing alcohol:

Non-dependent drinkers Brief GP intervention ↓ drinking in ~24%. Present results of screening interventions, e.g. AUDIT (➔ p. 159), and identify risks. Provide information about safe amounts of alcohol and harmful effects of exceeding these. Assess whether the patient is receptive to change. If so, agree targets to ↓ consumption, encourage, and negotiate follow-up.

Alcohol-dependent drinkers Suffer withdrawal symptoms if they ↓ alcohol consumption (e.g. anxiety, fits, delirium tremens—➔ p. 1103).

- If wanting to stop drinking—refer to the community alcohol team; suggest self-help organizations, e.g. Alcoholics Anonymous; involve family/friends
- Detoxification in the community usually uses a reducing regimen of chlordiazepoxide over a 1wk period. Various regimens are used e.g. 20–30mg qds on days 1 and 2; 15mg qds on days 3 and 4; 10mg qds on day 5; 10mg bd on day 6; 10mg od on day 7 then stop

If ambivalent/unwilling to change Provide information; reassess and re-inform on each subsequent meeting; support the family.

Vitamin B supplements People with chronic alcohol dependence are frequently deficient in vitamins, especially thiamine—give oral thiamine indefinitely (if severe, 200–300mg/d; if mild, 10–25mg/d). During detoxification in the community—give thiamine 200mg od for 5–7d.

Relapse Common. Warn patients; encourage to re-attend. Be supportive. Maintain contact (↓ frequency and severity of relapses). Consider drugs to prevent relapse, e.g. acamprosate, disulfiram (specialist initiation).

Delirium tremens ➔ p. 1103

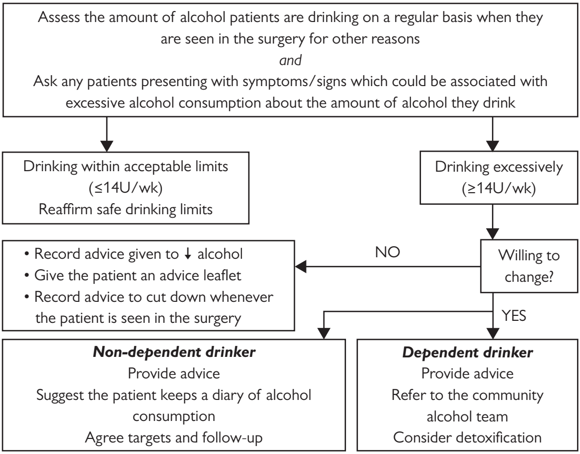

Assess the amount of alcohol patients are drinking on a regular basis when they are seen in the surgery for other reasons
and
Ask any patients presenting with symptoms/signs which could be associated with excessive alcohol consumption about the amount of alcohol they drink
Drinking within acceptable limits (≤14U/wk)
Reaffirm safe drinking limits
Drinking excessively (≥14U/wk)
Willing to change?
NO
YES
• Record advice given to ↓ alcohol
• Give the patient an advice leaflet
• Record advice to cut down whenever the patient is seen in the surgery
Non-dependent drinker
Provide advice
Suggest the patient keeps a diary of alcohol consumption
Agree targets and follow-up
Dependent drinker
Provide advice
Refer to the community alcohol team
Consider detoxification

Figure 7.7 Alcohol management strategy

⚠ Community detoxification is contraindicated for patients with:

- Poor home environment
- Poor cooperation
- Previous failed detoxification at home
- History of previously complicated withdrawal (e.g. withdrawal seizures or delirium tremens)
- ↑ risk of suicide
- Uncontrollable withdrawal symptoms
- Confusion or hallucinations
- Epilepsy or fits
- Malnourishment
- Severe vomiting/diarrhoea
- Acute physical/psychiatric illness
- Multiple substance misuse

Further information

DVLA Assessing fitness to drive—a guide for medical professionals. 🖱 www.gov.uk/government/publications/assessing-fitness-to-drive-a-guide-for-medical-professionals
NICE (2010, updated 2017) Alcohol use disorders: diagnosis and management of physical complications. 🖱 www.nice.org.uk/guidance/cg100
NICE (2010) Alcohol use disorders: prevention. 🖱 www.nice.org.uk/guidance/ph24
NICE (2011) Alcohol use disorders: diagnosis, assessment and management. 🖱 www.nice.org.uk/guidance/cg115
WHO Alcohol Use Disorders Identification Test (AUDIT): guidelines for use in primary care. 🖱 www.who.int/substance_abuse/publications/audit/en/
WHO Brief Intervention for Hazardous and Harmful Drinking: a manual for use in primary care. 🖱 www.who.int/substance_abuse/publications/audit_sbi/en/

Patient advice and support

ADFAM Support for families. 🖱 www.adfam.org.uk
Alcoholics Anonymous ☎ 0800 9177 650 🖱 www.alcoholics-anonymous.org.uk
Drinkline (government-sponsored helpline) ☎ 0300 123 1110

Assessment of drugs misuse

14% of ♂ and 8% of ♀ aged 16–59y report taking illicit drugs in the past year. The majority of patients on treatment programmes report opioid misuse (heroin—54%; methadone—13%), but the most frequently abused drugs are cannabis, amphetamine, ecstasy, and cocaine. Three factors appear important: availability of drugs; vulnerable personality; and social pressures—particularly from peers. Commonly misused drugs—Table 7.5, ➲ p. 164.

Controlled drugs regulations ➲ p. 124

Legal highs New psychoactive substances (NPS) ('legal highs', 'illegal legals', or 'illegal highs') are substances designed to produce similar effects to controlled drugs (e.g. cannabis, cocaine, and ecstasy), but structurally different enough to avoid being subject to the Misuse of Drugs Act. The UK Psychoactive Substances Act (2016) bans manufacture, import/export, or supply of NPS. Possession of NPS is not an offence.

Detection Warning signs suggesting drug misuse:

Use of services Suspicious requests for drugs of abuse (e.g. no clear medical indication, prescription requests are too frequent).

Signs and symptoms

- Inappropriate behaviour
- Lack of self-care
- Unexplained nasal discharge
- Unusually constricted/dilated pupils
- Evidence of injecting (e.g. marked veins)
- Hepatitis B/C or HIV infection

Social factors Family disruption, criminal history.

Assessment Assess on >1 occasion before deciding how to proceed. Exceptions are severe withdrawal symptoms and/or evidence of an established regimen requiring continuation. Points to cover:

General information

- Check identification (ask to see an official document)
- Contact with other agencies (including last GP)—check accuracy
- Current residence: family—partner, children
- Employment/finances
- Legal problems/criminal behaviour—past and present

History of drug use/risk-taking behaviour

- Reason for consulting now and willingness to change
- Current and past usage
- Unsafe sexual practices
- Knowledge of risks

Medical and psychiatric history

- Complications of drug abuse, e.g. HIV, hepatitis, accidents
- General medical and psychiatric history and examination
- Alcohol abuse
- Overdose—accidental/deliberate

Investigations

- Consider urine toxicology to confirm drug misuse
- Consider blood for FBC, LFTs, hepatitis B/C, and HIV serology (with consent and counselling—➲ p. 720), and other tests according to medical history/examination

- Specific drugs See Table 7.5, ➔ p. 164. ❗ Gabapentin/pregabalin are increasingly being used as drugs of abuse, particularly in prisons

Preventing prescription drug dependence

- Benzodiazepines and z-drugs ➔ p. 169
- Opioids ➔ p. 167

Prescribing for drug misusers Approach with special caution. Some controlled drugs can be dispensed to substance misusers in instalments providing they are prescribed on special NHS prescription forms (FP10 MDA—England; WP10 MDA—Wales; GP10—Scotland; HS21—Northern Ireland). The prescription specifies: number of instalments; intervals between instalments and, if necessary, instructions for supplies at weekends or bank holidays; total quantity of controlled drug providing treatment for a period ≤14d; and quantity to be supplied per instalment. As a general principle, substitute opioid medicines are prescribed in daily instalments. ❗ The prescription must be dispensed on the date on which it is due.

Other equipment for drug misusers Doctors, pharmacists, and drug workers may provide supplies of alcohol swabs, sterile water (≤10 ampoules of 2mL or less), mixing utensils, filters, and citric acid to drug misusers for the purposes of harm reduction.

Notification of drug misusers Patients who start treatment for drug misuse in the UK in specialized drug treatment centres have their details passed anonymously to national drug monitoring services. All types of problem drug misuse are reported. Databases cannot be used as a check on multiple prescribing as data are anonymized.

Driving and drugs misuse ➔ p. 99

Overdose ➔ p. 1098

Travelling abroad with controlled drugs ➔ p. 125

> ❗ The RCGP Substance Misuse Unit provides certificate courses in management of drug and alcohol misuse. 🖱 www.rcgp.org.uk

Further information

DH (2017) Drug misuse and dependence: UK guidelines on clinical management. 🖱 https://www.gov.uk/government/publications/drug-misuse-and-dependence-uk-guidelines-on-clinical-management

Advice and support for patients and their families

ADFAM Support for families 🖱 www.adfam.org.uk
Benzodiazepines 🖱 www.benzo.org.uk
Drugs-info 🖱 www.drugs-info.co.uk
Drugwise 🖱 www.drugwise.org.uk
Know the Score (Scotland) ☎ 0800 587 5879 🖱 www.knowthescore.info
Solvent abuse ☎ 01785 810762 🖱 www.re-solv.org
Talk to FRANK (England and Wales) Government-run information, advice, and referral service. ☎ (24h) 0300 123 6600 🖱 www.talktofrank.com

Table 7.5 Commonly misused substances in the UK

Name (street names)	How usually taken	Effects sought	Harmful effects
Heroin (smack, horse, gear, H, junk, brown, stag, scag, jack)	Injected, snorted, or smoked	Drowsiness, sense of warmth and well-being	Dependence/tolerance Overdose—can lead to coma and death Sharing injecting equipment brings risk of HIV/hepatitis infection
Cocaine (coke, charlie, snow, C)	Snorted in powder form, injected	Sense of well-being, alertness, and confidence	Dependence, restlessness, paranoia Damage to nasal membranes
Crack (freebase, rock, wash, stone)	Smokable form of cocaine	Similar to those of snorted cocaine but initial feelings are much more intense	As for cocaine but, because of the intensity of its effects, crack use can be extremely hard to control Additionally causes lung damage ('crack lung')
Ecstasy *(E, XTC, doves, disco biscuits, echoes, scooby doos)* *Chemical name: MDMA*	Swallowed, usually in tablet form	Alert and energetic, but with a calmness and a sense of well-being towards others. Heightened sense of sound and colour	Nausea and panic Overheating and dehydration—if dancing can be fatal Use has been linked to liver/kidney problems Long-term effects are not clear but may include mental illness and depression
LSD (acid, trips, tabs, dots, blotters, microdots)	Swallowed on a tiny square of paper	Hallucinations, including distorted/mixed-up sense of vision, hearing, and time. LSD trip can last 8–12h	There is no way of stopping a bad trip which may be a very frightening experience ↑ risk of accidents Can trigger long-term mental health problems
Magic mushrooms (shrooms, mushies)	Eaten raw or dried, cooked in food, brewed into tea	Similar effects to those of LSD, but the trip is often milder + shorter	As for LSD, with the additional risk of sickness and poisoning
Khat (quat, chat)	Chewed as leaves	Stimulant, ↓ sleep	Insomnia, irritability, panic

Barbiturates (barbs, downers)	Swallowed as tablets/capsules; injected	Calm and relaxed state, larger doses produce a drunken effect	Dependency/tolerance Overdose can lead to coma or death Severe withdrawal symptoms
Amphetamines (speed, whizz, uppers, billy, sulph, amp)	In powder form, dissolved in drinks, injected, sniffed or snorted	Stimulates the nervous system, wakefulness, feeling of energy and confidence	Insomnia, mood swings, irritability, panic The comedown (hangover) can be severe and last for several days
Cannabis (hash, dope, grass, blow, ganja, weed, shit, puff, marijuana)	Rolled with tobacco into a spliff, joint, or reefer and smoked; smoked in a pipe; or eaten	Relaxed, talkative state, heightened sense of sound and colour	Impaired coordination and ↑ risk of accidents Poor concentration, anxiety, depression ↑ risk of respiratory diseases, including lung cancer
Tranquillizers (include: Valium®, Ativan®, Mogadon® (moggies), temazepam (wobblies, mazzies, jellies))	Swallowed as tablets or capsules, injected	Prescribed for the relief of anxiety and to treat insomnia, high doses cause drowsiness	Dependency/tolerance ↑ risk of accidents Overdose can be fatal Severe withdrawal symptoms
Anabolic steroids (many trade names)	Injected or swallowed as tablets	With exercise, can help to build up muscle. Some debate about whether ↑ muscle power and athletic performance	♂: erection problems, risk of MI or liver disease ♀: development of male characteristics Injecting equipment: brings risk of HIV or hepatitis infection
Poppers (alkyl nitrates, including amyl nitrate with trade names such as Ram, TNT, Thrust)	Vapours from small bottle of liquid are breathed in through mouth or nose	Brief and intense head-rush caused by a sudden surge of blood through the brain	Hypotension/fainting Headaches Nausea Skin burns 'Sudden sniffing death syndrome'—due to arrhythmia
Solvents (including lighter gas refills, aerosols, glues). Some painter thinners and correcting fluids	Sniffed or breathed into the lungs	Short-lived effects similar to being drunk, thick-headed, dizziness, possible hallucinations	Nausea, blackouts, increased risk of accidents Fatal arrhythmias can cause instant death

Management of drugs misuse

Management aims to help the patient stay healthy until a drug-free life is achieved. In particular by ↓ the risk of infectious diseases, drug-related deaths, and criminal activity to finance drug habits. Care can be difficult due to chaotic lifestyle and drug-seeking behaviour. Set clear rules of engagement.

❗ GPs interested in working in specialist drug misuse treatment centres or providing substitute prescribing in the community should obtain specialist training. The RCGP Substance Misuse Unit provides certificate courses in management of drug and alcohol misuse (🖱 www.rcgp.org.uk).

Role of the GP

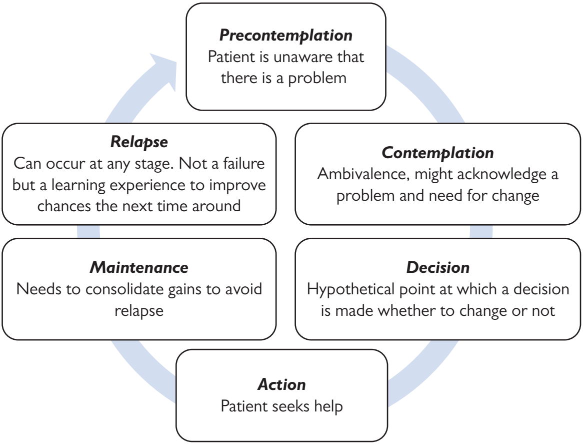
Important role in identifying drug misuse (including prescription drugs) and assessing willingness to modify drug behaviour (see Figure 7.8). GPs can also provide:

Information and advice as appropriate about:

- Safe injecting, overdose prevention and specific risks of drugs (Table 7.5, ➔ p. 164)—including local risks, e.g. contaminated street drugs
- Safe sexual practices (➔ p. 711)
- Driving and drug misuse (➔ p. 99)
- Other sources of support/information

Medical care

- Routine medical care, including treatment of complications of drug misuse, e.g. infected injection sites
- Routine preventive care (e.g. cervical screening, contraception)
- Blood-borne viruses (hepatitis B/C, HIV)—testing as needed; hepatitis B vaccination to injecting drug misusers not already infected/immune, and close contacts of those infected—use an accelerated regimen—immunization at 0, 7, and 21d, and a booster after 12mo



Precontemplation
Patient is unaware that there is a problem

Contemplation
Ambivalence, might acknowledge a problem and need for change

Decision
Hypothetical point at which a decision is made whether to change or not

Action
Patient seeks help

Maintenance
Needs to consolidate gains to avoid relapse

Relapse
Can occur at any stage. Not a failure but a learning experience to improve chances the next time around

Figure 7.8 Stages of change in addiction

- Referral for specialist assessment/treatment of drug misuse. ❗ Patients requiring substitute prescribing (e.g. with methadone/buprenorphine for heroin misuse) should be referred to a specialist treatment centre

Prescription benzodiazepine addiction ➔ p. 169

Prescription opioid misuse Suspect if taking long-term opioid medication for any non-cancer condition, particularly if drug-seeking behaviour (e.g. early requests/requests for replacement medication), refusal to see a GP for reviews, or reluctance/refusal to ↓ the dose of opioids.

Action Discuss your concerns with the patient directly. Offer patients the option of referral to a specialist drug treatment centre for substitute prescribing with methadone/buprenorphine, or detoxification.

Detoxification Convert patients on patches to long-acting oral medication. Agree a gradually ↓ dose of opioid, e.g. 10–25% of the dose/mo. ❗ Patients who detoxify should be warned of the risk of overdose if they relapse and restart opioids again at the same dose. Monitor frequently.

Opioid withdrawal symptoms

- Within 24h muscle aches; restlessness/anxiety; watering eyes/runny nose; excessive sweating; yawning; inability to sleep
- Later Diarrhoea; abdominal cramps; goosebumps ('cold turkey'); nausea/vomiting; blurred vision and dilated pupils; tachycardia; ↑ BP

Safe injecting advice Provide information about safer routes of drug administration, e.g. smoking/rectal administration for heroin abusers. Discourage IM/subcutaneous administration. Advise:

- Safe injecting—never inject alone; always inject with the blood flow; rotate sites—avoid neck, groin, penis, axilla, foot and hand veins, and any infected areas/swollen limbs. ❗ Poor veins indicate poor technique—find out what the patient is doing
- Drugs/equipment—sterile injecting equipment with small-bore needle; dispose of equipment safely after use; avoid unsuitable preparations, e.g. crushed tablets and/or drug cocktails
- First aid—learn basic principles of first aid/CPR; encourage calling for an ambulance. Suggest a naloxone kit (available from drug treatment centres)—↓ deaths from opioid overdose

Opioid overdose risk factors

- Injecting heroin
- Longer injecting career
- Depression, suicidal thoughts
- Recent non-fatal overdose
- High levels of use/intoxication
- High levels of alcohol use
- Lowered tolerance through detoxification/imprisonment
- Multiple drug use—particularly CNS depressants
- Sharing equipment/other high-risk injecting behaviour—may indicate low concern about personal risk
- Not being on/premature exit from a methadone treatment programme

Further information

DH (2017) Drug misuse and dependence: UK guidelines on clinical management. 🖱 https://www.gov.uk/government/publications/drug-misuse-and-dependence-uk-guidelines-on-clinical-management

Faculty of Pain Management Identification and treatment of prescription opioid dependent patients. 🖱 https://www.rcoa.ac.uk/faculty-of-pain-medicine/opioids-aware/clinical-use-of-opioids/identification-and-treatment

Insomnia

From the Latin meaning 'no sleep': describes a perception of disturbed or inadequate sleep. ~1:4 of the UK population (♀ > ♂) are thought to suffer in varying degrees. *Prevalence*: ↑ with age, rising to 1 in 2 amongst the over 65s.

Causes Numerous—common examples include:

- Minor, self-limiting—travel, stress, shift work, small children, arousal
- Psychological ~½ have mental health problems: depression, anxiety, mania, grief, alcoholism
- Physical—drugs (e.g. steroids), pain, pruritus, tinnitus, sweats (e.g. menopause), nocturia, asthma, obstructive sleep apnoea

Definition of 'a good night's sleep'

- <30min to fall asleep
- Maintenance of sleep for 6–8h
- <3 brief awakenings/night
- Feels well rested and refreshed on awakening

Management of insomnia

Careful evaluation. Many do not have a sleep problem themselves but a relative feels there is a problem, e.g. the retired milkman continuing to wake at 4 a.m. Others have unrealistic expectations, e.g. they need 12h sleep/d. Reassurance alone may be all that is needed.

For genuine problems with insomnia

- Eliminate physical problems preventing sleep, e.g. treat asthma/eczema; give long-acting painkillers to last the whole night; consider HRT or clonidine for sweats; refer if obstructive sleep apnoea is suspected (➔ p. 308)
- Treat psychiatric problems, e.g. depression, anxiety
- Sleep hygiene—see Box 7.2
- Relaxation techniques—podcasts; relaxation classes (often offered by local recreation centres/adult education centres); many physiotherapists can also teach relaxation techniques
- Consider drug treatment only as a last resort. Benzodiazepines should be prescribed for insomnia '*only when it is severe, disabling, or subjecting the individual to extreme distress*'

> ❗ Only prescribe a few weeks' supply at a time due to potential for dependence and abuse. Never put benzodiazepines or z-drugs on repeat prescription for sleeping problems. Beware the temporary resident who has 'forgotten' his/her night sedation.

Drug treatment Benzodiazepines (e.g. temazepam), zolpidem, zopiclone, and low-dose TCA (e.g. amitriptyline 10–75mg) nocte are all commonly prescribed for patients with insomnia. Side effects include amnesia and daytime somnolence. Most hypnotics do affect daytime performance and may cause falls in the elderly. Warn patients about their effect on driving and operating machinery

Box 7.2 Principles of 'sleep hygiene'

- Don't go to bed until you feel sleepy
- Don't stay in bed if you're not asleep
- Avoid daytime naps
- Establish a regular bedtime routine
- Reserve a room for sleep only (if possible). Do not eat, read, work, or watch TV in it
- Make sure the bedroom and bed are comfortable, and avoid extremes of noise and temperature
- Avoid caffeine, alcohol, and nicotine
- Have a warm bath and warm milky drink at bedtime
- Take regular exercise, but avoid late night hard exercise (sex is OK)
- Monitor your sleep with a sleep diary (record both the times you sleep and its quality)
- Rise at the same time every morning regardless of how long you've slept

Complications of insomnia

- Reduced quality of life
- Reduced concentration/memory—may affect performance of daytime tasks
- Relationship problems
- Risk of accidents—10% of motor accidents are related to tiredness

Obstructive sleep apnoea ➔ p. 308

Sleepwalking ➔ p. 891

Night terrors ➔ p. 891

Restless legs ➔ p. 552

Addiction to prescription benzodiazepines and z-drugs

Common in primary care. Regular use of hypnotics for insomnia rapidly leads to tolerance (in <1mo). Long-term use results in decline in cognitive functioning and blunting of emotions. In older people, hypnotic use is also associated with ↑ falls and associated injuries. However, stopping the drugs results in rebound insomnia and other effects which may be severe including anxiety, panic attacks, muscle pain, and rarely seizures.

Stopping hypnotics Discuss stopping hypnotics with any patient who has had regular prescriptions for >3mo. Explain long-term risks of taking these drugs. Switch to an equivalent dose of diazepam and agree a schedule for reduction—usually 1mg every 2wk, although can be slower if necessary. Withdrawal symptoms tend to occur 2–10d after reduction in dose.

Dose equivalence 5mg diazepam is approximately equivalent to 10mg temazepam, 5mg nitrazepam, 500mcg lorazepam, 15mg chlordiazepoxide, 15mg oxazepam, 7.5mg zopiclone, or 10mg zolpidem.

Patient information and support

Benzodiazepine addiction 🖱 www.benzo.org.uk
Royal College of Psychiatrists (2015) Sleeping well. 🖱 https://www.rcpsych.ac.uk/mental-health/problems-disorders/sleeping-well? www.rcpsych.ac.uk/healthadvice/problemsanddisorders/sleepingwell

Chapter 8

Chronic disease and elderly care

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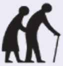
Maintaining independence *198*
Carers *200*

❗ In other sections of this book, where management differs from the norm for elderly patients, the text is highlighted in a box marked with this symbol.



Chronic disease management

The predominant disease pattern in the developed world is one of chronic or long-term illness. In the UK, 41% of adult ♂ and 43% of adult ♀ report a long-term illness. This figure is increasing as our population ages. People with long-term conditions are very intensive users of services; they account for 52% of GP appointments, 65% of outpatient appointments, and 77% of hospital bed days.

Long-term conditions frequently managed in general practice include:

- Back pain
- Cancer
- DM
- Dementia
- HIV
- Arthritis of all types
- Chronic lung disease
- Cardiovascular disease, e.g. ↑ BP, heart disease, stroke
- Renal or liver failure
- Irritable bowel syndrome
- Inflammatory bowel disease
- Chronic neurological conditions, e.g. Parkinson's disease, MS
- Psychiatric illness, e.g. depression, psychosis

Although details of chronic illness management depend on the illness, people with chronic diseases of all types have much in common with each other. They all have similar concerns/problems and must deal not only with their disease(s) but also its impact on their lives/emotions.

Common patient concerns

- Finding and using health services
- Finding and using other community resources
- Knowing how to recognize/respond to changes in a chronic disease
- Dealing with problems and emergencies
- Making decisions about when to seek medical help
- Using medicines and treatments effectively
- Knowing how to manage stress/depression that goes with chronic illness
- Coping with fatigue, pain, and sleep problems
- Getting enough exercise
- Maintaining good nutrition
- Working with your doctor(s) and other care providers
- Talking about your illness with family and friends
- Managing work, family, and social activities

Common elements of effective chronic illness management

- **Involvement of the whole family** Chronic diseases do not only affect the patient but everyone in a family
- **Collaboration between service providers, patients, and carers** Negotiate and agree a definition of the problem; agree targets and goals for management; develop an individualized self-management plan
- **Personalized written care plan** Take into account patients'/carers' views and experience and the current evidence base
- **Tailored education in self-management** A patient with diabetes spends ~ 3h/y with a health professional—the other 8757h he or she manages his/her own condition. Helping patients with chronic disease understand and take responsibility for their conditions is vital
- **Planned follow-up** Proactive follow-up according to the care plan—use of disease registers and call–recall systems is important

- **Monitoring of outcome and adherence to treatment** Use of disease/treatment markers; monitoring of concordance, e.g. checking prescription frequency; medicine management programmes—➔ p. 116
- **Tools and protocols for stepped care** Provide a framework for using limited resources to greatest effect; step professional care in intensity—start with limited professional input and systematic monitoring, then augment care for patients not achieving an acceptable outcome
- **Targeted use of specialist services** For those patients who cannot be managed in primary care alone
- **Monitoring of process** Continually monitor management through clinical governance mechanisms (➔ p. 52)

Multimorbidity ➔ p. 174

Depression and chronic disease^{N} Depression is common among people with chronic disease. It is reported to affect 30–50% of those with epilepsy, CVD, dementia, cancer, type 2 DM, and arthritis.

Interaction between depression and chronic physical illness Depression in those with chronic medical illnesses adversely affects prognosis. Conversely, treatment of depression can improve prognosis.

Depression is associated with:
- ↑ mortality, ↑ morbidity, ↑ disability, and poorer quality of life
- ↑ prevalence of smoking and sedentary lifestyles
- Poorer chronic disease outcome measures, e.g. higher HbA1c levels
- ↑ use of services and ↑ healthcare costs
- Poor concordance with medication and management plans

Detection of depression Use NICE depression screening questions:
- During the last month, have you often been bothered by feeling down, depressed, or hopeless?
- During the last month, have you often been bothered by having little interest or pleasure in doing things?

A positive response to either of these questions should prompt further assessment with the following 3 questions: *During the last month have you often been bothered by*:

- Feelings of worthlessness?
- Poor concentration?
- Thoughts of death?

Further assessment and management ➔ p. 978

Residential care homes 220,000 people live in residential care in England; 77% are elderly with long-term health conditions. Of younger residents, 61% have learning disabilities, 21% have mental illness, 17% have physical disability, and 2% have substance misuse problems. Chronic disease management for residents of care homes is frequently neglected. Ensure that chronic disease management checks take place including routine blood monitoring, physical health checks, and medication review.

Deprivation of Liberty Safeguards (DoLS) ➔ p. 1106

Further information

NICE (2009) Depression in adults with a chronic physical health problem.
🔗 www.nice.org.uk/guidance/cg91

Multimorbidity

Definition and statistics Multimorbidity refers to the presence of ≥2 long-term health conditions in a single individual. It is common: 1 in 6 patients in the UK has >1 QOF condition; 65% of people >65y and 82% of those aged >85y have ≥2 conditions (♀ > ♂). However, although prevalence of multimorbidity ↑ with age, multimorbidity can affect all age groups. It is much more common in socially deprived populations and complexity tends to increase the more conditions a patient has.

Challenges of multimorbidity

Single disease guidelines Since the early 1990s, a plethora of guidelines have been produced synthesizing research around single disease entities into 'best practice' that we have learned to follow rigidly. However, most research is carried out on populations without co-morbidities, so may not be applicable to patients with multimorbidity. Rigid adherence to guidelines for patients with multimorbidity can lead to polypharmacy with subsequent adverse effects, and conflicting treatment targets/advice.

Condition overlap Some conditions have similar symptoms and signs, resulting in diagnostic overlap. For example, breathlessness may be due to heart failure or COPD. One condition may also have an impact on another. For example, chronic pain can result in worsening of depression. This makes unravelling a patient's medical problems very difficult.

Polypharmacy It is easy to keep adding medication as new guidelines are released, to reach performance targets or as patients present with symptoms. It is much harder to stop medication. The more medication a patient is taking, the greater the risk of interactions between medications and adverse effects—➔ p. 194.

Medical complexity In a traditional GP appointment, the GP works alone and has 10min to assess the patient and formulate a management plan. That is simply not enough time for highly complex patients with multiple interacting and interdependent problems, requiring a holistic multidisciplinary approach. Time pressure can result in inadequate assessment and/or disengagement of the patient/carers.

High use of services Patients with multimorbidity use more GP appointments, take more medication, have more outpatient hospital appointments, and are likely to be admitted to hospital both for routine and emergency reasons.

Disorganization/fragmentation of care The medical model of care uses a systems-based approach. Early specialization of doctors has resulted in silo working. Patients with multimorbidity are frequently under review by several different specialist medical teams resulting in mixed messages and prioritization of some of their problems over others, regardless of whether these are priorities for the patient. Even in primary care, it is common for patients with multimorbidity to be asked to attend multiple chronic disease reviews in the same practice.

Patient burden Multimorbidity is a huge burden for patients and carers. They may have to attend multiple appointments and manage complicated drug regimens as well as dealing with the effects of their illnesses and side effects of treatment. Multimorbidity is associated with poorer quality of life and ↑ risk of anxiety and depression.

Identifying patients at risk of adverse events[N] Not all patients with ≥2 conditions need additional support. As multimorbidity is common, it is important to focus resources on those who are most at risk of adverse events. Consider:

- Patients at high risk of unplanned/care home admission Computer algorithms aid identification e.g. Electronic Frailty Index (eFI), QAdmissions, Predicting Emergency Admissions Over the Next Year (PEONY)
- Number of regular prescribed medications In general, patients taking >15 regular prescribed medications are at high risk; those taking 10–14 medications are at moderate risk
- Frailty (➔ p. 190) Assess using: gait speed (>5sec to walk 4m), self-reported health status on a 0–10 scale (<6 suggests frailty), or formally validated tools, e.g. PRISMA-7 questionnaire (a score of ≥3 suggests ↑ risk of frailty and the need for further review)

Holistic approach to management of multimorbidity

- For patients at high risk of adverse events, consider longer review appointments and try to ensure continuity, e.g. with a named GP
- Coordinate care in the practice, e.g. routine blood tests all done at the same time; all routine chronic disease reviews during the same appointment; quantities of repeat medicines and review dates aligned
- List conditions and problems from the patient's/carer's perspective
- Explore how the person's health conditions and treatments interact and how this affects quality of life
- Discuss the patient's goals, health priorities, and treatment preferences—this may also include advance care planning decisions (➔ p. 97)
- Discuss benefits and risks of following recommendations from guidance on single health conditions; review medications/treatments weighing risks vs benefits of each with the patient
- Explore ways to ↑ quality of life by ↓ treatment burden, adverse events, and unplanned care; focus on solving problems and symptom control; be alert for depression—consider screening (➔ p. 173)
- Create a care plan and share this across the multidisciplinary team highlighting the person responsible for coordination of care; ensure that the patient and carers understand the plan and know what to do if anything changes and in emergency situations
- Review the patient and care plan regularly

Further information

Cassell A, et al. (2018) The epidemiology of multimorbidity in primary care: a retrospective cohort study. *BJGP* **68**:e245–51.
NICE (2016) Multimorbidity: clinical assessment and management. 🖱 www.nice.org.uk/guidance/ng56
PRISMA-7 questionnaire 🖱 www.cgakit.com/fr-1-prisma-7
Wallace E, et al. (2015) Managing patients with multimorbidity in primary care. *BMJ* **350**:h176.

Genetics and genomics

There are 46 chromosomes—22 matching pairs with matching genes (autosomes) and 1 pair of sex chromosomes which may match (XX—♀) or differ (XY—♂). Genetics refers to the study of individual genes. Genomics refers to a person's entire genetic code—their genome. There are many ways in which variations in our genetic make-up result in disease.

Changes in chromosome number

- **Alteration in number of chromosomes** e.g. Down's syndrome (extra chromosome 21)
- **Sex chromosome abnormalities**—a sex chromosome is duplicated or deleted, e.g. Turner's syndrome (XO); Klinefelter's syndrome (XXY or XXYY)

Gross structural changes in chromosomes

- **Translocation** Part of one chromosome is transposed or translocated onto another. If no genetic information is lost there is no clinical effect (balanced translocation), although offspring often have problems. 6% of children with Down's syndrome have a translocation
- **Deletion** Loss of a portion of chromosome, e.g. cri-du-chat syndrome (deletion of the short arm of chromosome 5)

Single gene abnormalities

Autosomal dominant inheritance (Figure 8.1) >1000 diseases all individually rare. Heterozygotes have the disease; 1 in 2 pregnancies of an affected individual are affected—usually ♂ = ♀. Expression of the gene may vary, e.g. tuberous sclerosis (➔ p. 840); Marfan's syndrome (➔ p. 253); myotonic dystrophy (➔ p. 550); neurofibromatosis (➔ p. 552). Occasionally genes may be co-dominant and both the gene inherited from the mother and the gene inherited from the father are expressed simultaneously, e.g. blood types.

Autosomal recessive inheritance (Figure 8.2) >700 diseases that only manifest in the homozygote. Heterozygotes may be asymptomatic or have milder abnormalities. To develop severe disease, the affected gene must be inherited from both parents. The risk of an affected pregnancy is 1 in 4—usually ♂ = ♀. Affected individuals have unaffected children unless their partner is a heterozygote *Examples*: sickle cell disease (➔ p. 645); thalassaemia (➔ p. 644); cystic fibrosis (➔ p. 300).

Sex-linked disorders (Figure 8.3) ~100 are recognized. Most are recessively inherited from the mother and affect only ♂ offspring. A ♂ child of a heterozygote mother has a 1 in 2 chance of developing the disease; a ♀ child has a 1 in 2 chance of being a carrier. *Examples*: fragile X syndrome (➔ p. 840); haemophilia (➔ p. 646), red-green colour blindness (➔ p. 948), Duchenne's muscular dystrophy (➔ p. 550).

Mitochondrial disorders There are many thousands of mitochondria in each cell. Mitochondria have their own DNA (mtDNA). Mitochondrial conditions affect ♂ and ♀ equally. They result from mutations in mtDNA which can only be inherited from an affected ♀, and so are always passed through the female family line. *Example*: Leber's optic atrophy.

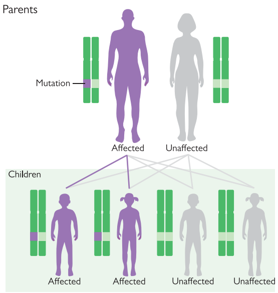

Parents
Mutation
Affected
Unaffected
Children
Affected
Affected
Unaffected
Unaffected

Figure 8.1 Autosomal dominant inheritance

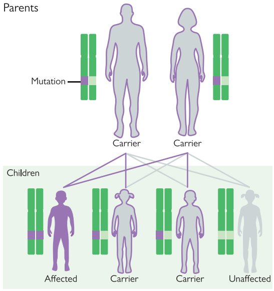

Parents
Mutation
Carrier
Carrier
Children
Affected
Carrier
Carrier
Unaffected

Figure 8.2 Autosomal recessive inheritance

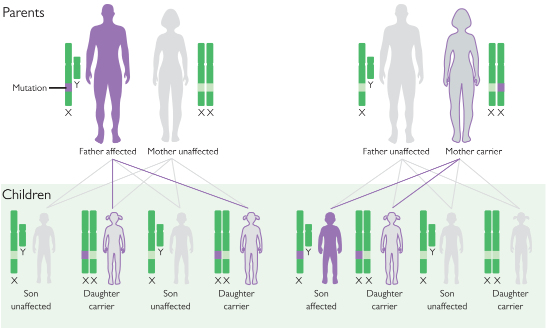

Parents
Mutation
Y
X
X X
Father affected
Mother unaffected
Y
X
X X
Father unaffected
Mother carrier
Children
Son unaffected
Daughter carrier
Son unaffected
Daughter carrier
Son affected
Daughter carrier
Son unaffected
Daughter carrier

Figure 8.3 Sex-linked recessive inheritance

Illustrations courtesy of National Library of Medicine (US). Genetics Home Reference. https://ghr.nlm.nih.gov/gallery

New mutations Not all who present with a genetic condition have a family history. Often, a new genetic mutation will arise either when the egg/sperm is formed, or at the embryonic stage.

Polygenic inheritance *All* diseases result from a combination of genetic and environmental factors. Variations in different parts of an individual's genome might ↑ or ↓ the chances of developing many common diseases such as cancer, diabetes, or heart disease.

Cancer genes (oncogenes) Alterations in the genetic make-up of cancer cells. Usually control cell division/multiplication. Not inherited. Identification of faulty tumour genes can potentially facilitate choice of cancer treatment and improve effectiveness/prognosis.

Personalized medicine This is a move away from a 'one-size-fits-all' approach to management of disease. Recent advances in genomic medicine (both in terms of testing and mapping of the human genome) now enable us to integrate information from the individual's genomic code together with the patient's history to help us better identify those at risk of disease, and target management options to improve treatment outcomes.

Medically unexplained symptoms

Medically unexplained symptoms (MUS) are physical symptoms for which no organic cause can be demonstrated. GPs deal with MUS in about 25% of consultations and MUS cost the NHS £3.1 billion/y. MUS cause disability as severe as that caused by illness with medically explained causes.

Epidemiology MUS are common throughout the world in all ages. Risk factors for development of MUS include:

- ♀ > ♂
- Physical illness/trauma
- Stressful life events, e.g. illness or death of a close relative, domestic violence, history of child abuse
- Media campaigns that highlight specific diseases

Classification MUS can be divided into 3 types of complaint:

- Pain of a specific location, e.g. back pain, headache, fibromyalgia
- Functional disturbance in a particular organ, e.g. IBS, palpitations
- Fatigue/exhaustion, e.g. chronic fatigue syndrome

Many patients have >1 MUS. There are also common overlaps of symptoms, e.g. patients with IBS often meet diagnostic criteria for chronic pelvic pain and vice versa.

Concurrent psychological illness 30% of patients with MUS have an underlying psychiatric problem—usually anxiety or depression.

Underlying mechanism MUS may be caused by a clear trigger, e.g. an illness or chemical exposure, but 2 mechanisms seem to underpin MUS:

- **Enhanced sense of bodily awareness** Tendency to notice and amplify normal physical sensations such as heartbeat. Over-awareness ↑ anxiety which, in turn, makes the bodily sensation more likely
- **Misattribution of symptoms** Rather than normalizing symptoms (e.g. *I have a headache because I've been working too hard*), patients with MUS tend to attribute somatic explanations (e.g. *I have a headache because I have a brain tumour*)

Assessment Consider a diagnosis of MUS in any patient with physical symptoms for >3mo that are affecting functioning but cannot be readily explained. Even if the patient is known to present with MUS, perform your assessment without prejudice. Patients with MUS have the same chance of developing serious new illnesses as any other patient. *Ask*:

- What are the symptoms? Rule out 'red flags'
- How much and what type of impairment do the symptoms cause?
- What are the patient's concerns about the symptom? Has the patient sought information from other sources e.g. Internet, friends?
- What made the patient come to the surgery today?
- What would the patient like you to do for him/her?
- Are there any signs of disease on physical examination?
- Does the patient have low mood or any symptoms of anxiety? Consider depression and/or anxiety screening questionnaires
- Are there any other social/psychological factors that may be triggering symptoms? e.g. family member or close friend who is ill; domestic violence; debt; work problems; past history of child abuse

Investigation Review patient notes carefully before requesting investigations. Usually investigations are used to clarify diagnosis and reassure the patient and GP. However, in patients with MUS:

- >50% of patients are not reassured following negative investigations
- False-positive results lead to ↑ anxiety and further investigation
- Colluding with the patient ↑ illness behaviour

❗ Find a balance between appropriate investigation and risk of harm through over-investigation. Prior to doing investigations, explain why they are being done and the meaning of negative results.

Management[G] 4 key areas:

- **Connecting** Go back to the beginning, listen to the patient, acknowledge suffering, use existing knowledge of the individual (or recognize that you have no knowledge of the individual)
- **Summarizing** Allow the patient to summarize problems, recap your understanding of the problem to the patient, give an explanation, and show your interest in the problem
- **Hand over** Develop a shared action plan or personal health plan with realistic goals to improve functioning, and provide reassurance about long-term outcome
- **Safety netting** Share uncertainty, inform patients about red flags indicating serious disease, and offer access should symptoms change

Regular appointments may be helpful, as may a brief physical examination at each visit to check for signs of disease. Offering suggestions for self-management (e.g. doing voluntary work, increasing physical activity levels) can be useful. Avoid referral unless there is a clear medical indication.

General treatment options If self-help is ineffective, try:

- **Antidepressant medication** e.g. amitriptyline 10mg at 5 p.m. (may be unlicensed). As response is often not dose dependent, start with a low dose. Explain that the drug is not being used to treat depression
- **CBT** Allows patients to develop changes in thinking/behaviour that will help them cope more effectively with their problems

Management of specific MUS

- Fibromyalgia—➔ p. 504
- IBS—➔ p. 388
- Chronic fatigue—➔ p. 502
- Atypical facial pain—➔ p. 531
- Chronic pelvic pain—➔ p.690
- Interstitial cystitis—➔ p. 423
- Tension-type headache—➔ p. 530
- TMJ dysfunction—➔ p. 910–11
- Globus—➔ p. 356
- Somatization disorder—➔ p. 975

Work Encourage patients to work if possible—➔ p. 92

Prognosis 4–10% of patients with MUS go on to have an alternative organic explanation; of those with true MUS, 25% will have ongoing symptoms after 12mo.

Somatization disorder ➔ p. 975

Further information

RCGP/Royal College of Psychiatrists/Trailblazers/National Mental Health Development Unit (2011) Guidance for health professionals on MUS. 🖰 https://www.rcpsych.ac.uk/pdf/CHECKED%20MUS%20Guidance_A4_4pp_6.pdf

Assessment of pain

Take a history to ascertain:

- What the patient means when he or she complains of pain
- The cause of the pain
- The severity of the pain

❗ Do not jump to conclusions/make assumptions about a patient's pain.

Assessment questions There are many approaches to assessing pain. The specifics of each scheme are not crucial—but it is important the scheme used has a logical outline which works for the individual clinician. A simple mnemonic approach is detailed in Figure 8.4. Always note any past history of addiction to prescription or illicit drugs.

Elderly patients and patients with difficulty communicating High prevalence of pain in the elderly population is now well recognized. 40–80% of elderly people in institutions are in pain. The reason for this lies in the difficulty in assessing those with communication difficulties. Additionally, the elderly often minimize their pain making it even more difficult to evaluate.

Methods of evaluation Unusual behaviour and its return to normal with adequate analgesia, may be the only confirmation of pain in patients with communication difficulties. Examples include:

Verbal expression e.g.

- Crying when touched
- Shouting
- Becoming very quiet
- Swearing
- Grunting
- Talking without making sense

Behavioural expression e.g.

- Jumping on touch
- Hand pointing to body area
- Increasing confusion
- Rocking/shaking
- Not eating
- Staying in bed/chair
- Grumpy mood

Facial expression, e.g.

- Grimacing/wincing
- Closing eyes
- Worried expression
- Withdrawn/no expression

Physical expression, e.g.

- Cold
- Pale
- Clammy
- Change in colour
- Change in vital signs if acute pain (e.g. BP, pulse)

Pain assessment tools Sometimes it is helpful to use pain scales to assess the degree of pain that a patient is in—particularly if communication is difficult. The most commonly used tool is a simple visual analogue pain scale—this consists of a line marked in graduations from 0 to 10. Ask patients to point to the place on the line which represents how much pain they are in, where 10 is the most possible pain and 0 is no pain.

Examine the patient The cause of the problem may be clear to you from history alone but examine the patient to confirm/refute your proposed diagnosis.

S	*Site of pain* Where? Any radiation? Numbness where pain felt? Pattern of involvement?
O	*Onset* When did it start? How did it start? What started it? Change over time?
C	*Character of pain* Type of pain—burning, shooting, stabbing, dull, etc.; pattern of pain, e.g. colicky, constant, etc.
R	*Radiation* Does the pain go anywhere else?
A	*Associated features* Are there any skin or joint changes, e.g. bruising, redness, or swelling?
T	*Timing/pattern* Is it worse at any time of day? Is it associated with any particular activities, e.g. movement, urination, eating, passing stool, coughing?
E	*Exacerbating and relieving factors*
S	*Severity* Record, especially if the pain is chronic and you want to measure change over time. Consider a patient diary. *Ask about:* • Pain intensity, e.g. none–mild–moderate–severe; rank on a 1–10 scale • Record interference with sleep or usual activities • Pain relief, e.g. none–slight–moderate–good–complete

Figure 8.4 Points to consider when taking a history of pain

⚠ Beware of emergency requests for opioids from patients unknown to you or your practice.

Further information

Livewell with Pain 🖱 https://livewellwithpain.co.uk/
Schofield P (2018) The assessment of pain in older people: UK national guidelines. *Age Ageing* **47**:1–i22.

Patient support

Action on Pain ☎ 0345 603 1593 🖱 www.action-on-pain.co.uk
Pain Association of Scotland ☎ 0800 783 6059 🖱 www.painassociation.com
Pain Concern ☎ 0300 123 0789 🖱 www.painconcern.org.uk

Principles of pain control

Acute pain Symptom of injured/diseased tissue. Subsides as the injury heals. Can be worsened by fear. Treat the underlying cause.

Chronic pain Defined as pain persisting for >3–6mo. Affects ~7% of adults in the UK. Cause is often multidimensional—with physical, social, and psychological factors all contributing to the overall feeling of pain.

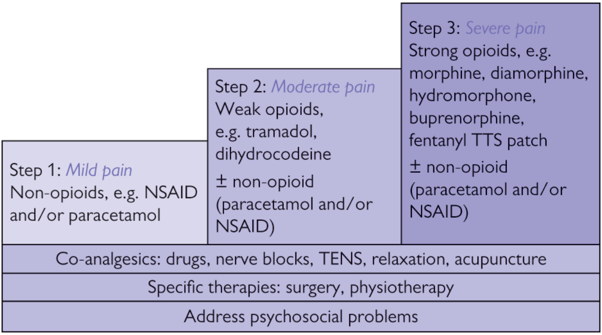
Goals of chronic pain management

- Set realistic targets—abolition of pain may be impossible—70% have pain despite analgesia
- If analgesia is not helping—stop it
- The aim is often rehabilitation with ↓ in distress/disability

Strategies for pain management A multidisciplinary approach is essential. *Consider*:

- Prevention e.g. wrist splints for carpal tunnel syndrome; analgesia prior to minor surgery
- Removal of cause Treat medical causes of pain e.g. infection, ↓ blood sugar (diabetic neuropathy). Refer surgical causes for surgery if surgery is appropriate, e.g. hip OA—joint replacement
- Pain-relieving drugs Start with a single drug at low dose and step up dose or add another drug as needed. Especially in situations of acute pain, step down if pain diminishes
- Physical therapies Acupuncture, physiotherapy, or TENS
- Nerve blocks Consider referral for epidural (low back pain), local nerve block, or sympathectomy (e.g. vascular rest pain)
- Modification of emotional response Psychotropic drugs, e.g. anxiolytics, antidepressants
- Modification of behavioural response e.g. back pain—consider referral to a back rehabilitation scheme

WHO analgesic ladder Use a step-by-step approach (Figure 8.5).



Step 1: *Mild pain*
Non-opioids, e.g. NSAID and/or paracetamol

Step 2: *Moderate pain*
Weak opioids, e.g. tramadol, dihydrocodeine
± non-opioid (paracetamol and/or NSAID)

Step 3: *Severe pain*
Strong opioids, e.g. morphine, diamorphine, hydromorphone, buprenorphine, fentanyl TTS patch
± non-opioid (paracetamol and/or NSAID)

Co-analgesics: drugs, nerve blocks, TENS, relaxation, acupuncture

Specific therapies: surgery, physiotherapy

Address psychosocial problems

Figure 8.5 World Health Organization (WHO) analgesic ladder

Source: World Health Organization (WHO). *Cancer Pain Relief*. Geneva, Switzerland: World Health Organization. Copyright © WHO 1986. https://www.who.int/cancer/palliative/painladder/en/

Step 1: non-opioid Start treatment with paracetamol. Stress the need for *regular* dosage. Adult dose is 1g every 4–6h (maximum daily dose 4g). If this is not adequate in 24h, either try a NSAID, e.g. ibuprofen 400mg tds (if appropriate), alone or in combination with paracetamol, or proceed to step 2.

Step 2: weak opioid + non-opioid Start treatment with a combined preparation of paracetamol + codeine/dihydrocodeine. Combining 2 analgesics with different mechanisms of action enables better pain control than using either alone. Combinations have ↓ dose-related side effects but the range of side effects is ↑ (additive effects of 2 drugs). Combinations using 30mg of codeine are more effective than paracetamol alone but it is cheaper and more flexible if constituents are prescribed separately, e.g. 'paracetamol 500mg/codeine 30mg'. Advise patients to take tablets regularly and not to assess efficacy after only a couple of doses.

❗ There is no proven additional analgesic benefit for preparations containing paracetamol + 8mg of codeine, compared to paracetamol alone.

Step 3: strong opioid + non-opioid

- Use immediate-release morphine tablets or morphine solution. 2 tablets of co-codamol 500/30 contain 60mg of codeine which is equi-analgesic to ~6mg of oral morphine. If changing to morphine, use a minimum dose of 5mg (6mg is hard to prescribe)
- Chronic pain may not be opioid sensitive. Give for a 1–2wk trial and *only* continue if of proven benefit. If the pain seems responsive to opioids and there are no undue side effects, ↑ the dose upwards by 30–50% every 24h until pain is controlled to a maximum of 120mg/d morphine equivalent—➲ p. 186

⚠ Take care if the patient is elderly or in renal failure—consider starting with a ↓ dose of morphine.

Addition of co-analgesics and adjuvant drugs In combination with analgesics, can enhance pain control. Examples include:

- Antidepressants—in low dose for nerve pain and sleep disturbance associated with pain; in larger doses for secondary depression
- Anticonvulsants—neuropathic pain
- Corticosteroids—pain due to oedema
- Muscle relaxants—muscle cramp pain
- Antispasmodics—bowel colic
- Antibiotics—infection pain
- Night sedatives—when lack of sleep is lowering pain threshold
- Anxiolytics—when anxiety is making pain worse (relaxation exercises may also help in these circumstances)

Referral If unable to remove cause and unable to achieve adequate pain relief, consider referral to a specialist pain control clinic or palliative care (depending on the context of the pain).

⚠ Be aware of secondary gain from pain if symptoms seem out of proportion (outstanding compensation claims are a significant negative factor in success of pain management).

Pain-relieving drugs

Paracetamol As effective a painkiller as ibuprofen. No anti-inflammatory effect but potent antipyretic. Drug of choice in OA where inflammation is absent. Side effects are rare. Overdose (>4g/24 h) can be fatal causing hepatic damage sometimes not apparent for 4–6d. Inadvertent overdose is easy due to presence of paracetamol in most OTC cold preparations—if suspected, refer immediately to A&E.

Non-steroidal anti-inflammatories (NSAIDs) (Table 8.1) Anti-inflammatory, analgesic, antipyretic. Start at the lowest recommended dose and do not use >1 NSAID concurrently. 60% respond to any NSAID—for those who don't, another may work. Selective inhibitors of cyclo-oxygenase-2 (COX2) have lower GI side effects, but should not be given to any patient with pre-existing, or high risk of, CVD.

Table 8.1 Commonly used NSAIDs

Drug	Dosage	Features
Ibuprofen	1.2–1.8g/d in 3–4 divided doses	Fewer side effects than other NSAIDs. Anti-inflammatory properties are weaker. Do not use if inflammation is prominent, e.g. gout. Higher doses (>1.2g/d) are associated with ↑ risk of thrombotic events and MI
Naproxen	0.5–1g/d in 1–2 divided doses	Good efficacy with a low incidence of side effects. Associated with lower thrombotic risk than other NSAIDs
Diclofenac	75–150mg/d in 1–2 divided doses	Good efficacy with a low incidence of side effects. Associated with ↑ thrombotic risk and ↑ liver reactions (drug-induced hepatitis) compared to other NSAIDs

Common side effects

- GI Associated with intestinal ulceration ± GI bleeding. GI side effects are more common in the elderly, people with liver disease or who have high alcohol intake, and those taking continuous low-dose aspirin, steroids, or SSRIs—avoid if possible. Warn all patients about the risk of GI side effects when taking NSAIDs and advise them to always take NSAIDs after food. If at high risk, >50y, or taking long-term NSAIDs, always co-prescribe stomach protection, e.g. with a PPI. NSAIDs may also cause exacerbation of pre-existing Crohn's disease or UC
- Sodium and water retention This can lead to deterioration in renal function ± renal failure. Avoid NSAIDs if possible in patients with renal impairment, liver disease or high alcohol consumption, hypertension, and/or heart failure. Where prescribing is unavoidable, monitor renal function while taking NSAIDs
- Impairment of ♀ fertility Avoid in women trying to conceive

Topical NSAIDs Of proven benefit for acute and chronic conditions and can be as effective as oral preparations. They have lower incidence of GI and other side effects, although these still can occur.

Use of opioids Useful for management of acute pain (e.g. following acute injury) and cancer pain, especially at the end of life. Little evidence they are helpful for long-term pain. In the UK, 8–12% of long-term prescribed opioid users meet criteria for current or past opioid use disorder.

General rules for initiating opioid therapy

- Explain that opioids are most effective for the treatment of acute pain; for the majority (>70%) opioids are not effective for long-term pain which requires other strategies
- Stress the aim of treatment is *not* complete abolition of pain, but ↓ in pain to allow the patient to use other strategies for symptom relief
- Warn about adverse effects including effects on driving
- Prescribe initially for a trial period only (1–2wk) and set defined outcomes in terms of pain, functional ability, and sleep (if impaired by pain)
- Set rules for stopping the drug—e.g. if outcomes are not achieved in 1–2wk, if the underlying condition resolves, if there are intolerable side effects, or if the patient is found to have abused the medication
- Explain that no further prescriptions will be issued if the patient does not come for review following the opioid trial

Management of prescription opioid misuse ➔ p. 167

Weak opioids Commonly used in primary care include codeine, dihydrocodeine, and tramadol.

Codeine and dihydrocodeine The most commonly used weak opioids in the UK. Standard adult dose is 30–60mg every 4h to a maximum of 240mg/24h. Analgesic effect is ↑ by regular ingestion. 10mg of codeine/dihydrocodeine equipotent to ~1mg of morphine. Effects of codeine/dihydrocodeine are reduced by concurrent use of antipsychotics (e.g. chlorpromazine, haloperidol), tricyclic antidepressants (e.g. amitriptyline), and metoclopramide. 5–10% of Caucasians have CYP2D6 genotype and lack a hepatic enzyme needed to convert codeine to morphine. They gain less analgesic effect codeine/dihydrocodeine.

Tramadol A synthetic analogue of codeine. Standard adult dose is up to 400mg/24h. It produces analgesia by 2 mechanisms: an opioid effect, and an enhancement of serotonergic and adrenergic pathways. Compared to codeine/dihydrocodeine, oral tramadol is absorbed faster giving analgesia in <1h with peak action in 1–2h. Tramadol is also metabolized in the liver, so safer for the elderly and those with renal impairment, as associated with fewer opioid side effects (notably less respiratory depression and constipation). However, nausea and vomiting can be a problem and rarely tramadol can trigger psychiatric reactions.

❗ Always consider co-prescribing a laxative to any patient taking opioids.

Opioid side effects ➔ p. 186

Morphine and other strong opioids ➔ p. 186

Further information

Faculty of Pain Medicine Opioids Aware. 🖱 www.rcoa.ac.uk/faculty-of-pain-medicine/opioids-aware
Livewell with Pain 🖱 https://livewellwithpain.co.uk/

Morphine and other strong opioids

Morphine is the strong opioid of first choice for moderate to severe pain in both malignant and non-malignant conditions.

General rules for initiating opioid therapy ➲ p. 185

Starting oral morphine Start with 4-hourly immediate release morphine. Give clear instructions. *Initial dosage*:

- Adults not pain-controlled with regular weak opioids, 5–10mg every 4h
- Elderly or cachectic patients, or those not taking regular weak opioids 5mg every 4h (2.5mg if very elderly/frail)

Titration of dose ↑ dose as needed by 25–50%/d until pain is controlled, or the patient experiences unacceptable side effects, to a maximum of 120mg/d. ⚠ Risk of harm increases substantially at doses >120mg/d with no ↑ benefit.

❗ Never attempt dose titration for unstable pain using a fentanyl patch—convert from oral morphine once a stable dose is attained.

Maintenance Once pain is controlled, consider a long-acting preparation of equivalent dose (e.g. MST® bd, MXL® od). Calculate total daily dose of morphine by adding together the 4-hourly doses.

Increasing dose If further increase in dose is necessary, use a 25–50% dose increment to a maximum of 120mg/d. ↑ dose rather than frequency of administration—slow-release tablets are designed for od/bd dosing.

Breakthrough pain Pain of rapid onset, and moderate/severe intensity despite background analgesia. *Management*:

- Prescribe immediate release morphine for breakthrough pain—give the equivalent 4-hourly dose as an additional dose
- If pain starts to occur regularly before the next dose of analgesia is due, consider ↑ the regular background dose

Common side effects of opioid drugs Warn patients:

- Nausea/vomiting—affects >1 in 3 patients for the first 2wk of use. Prescribe a regular antiemetic for 2wk, e.g. cyclizine 50mg tds. If nausea/vomiting continues, consider an alternative opioid
- Constipation—consider prescribing prophylactic laxatives, e.g. bisacodyl 1–2 tab nocte. Fentanyl causes less constipation than morphine
- Drowsiness/cognitive impairment—usually improves within the first week. Advise patients not to drive, perform other skilled tasks, or work with dangerous machinery if affected. If not improving, consider an alternative opioid, or refer for specialist advice

Conversions to other preparations See Table 8.2

Reasons to choose/switch to an alternative strong opioid

- Unacceptable side effects
- Renal failure—fentanyl is licensed for patients with renal failure; oxycodone is safe in mild/moderate renal failure
- Unable to take oral medication regularly—consider fentanyl or buprenorphine patch, or syringe driver

Controlled drug prescriptions ➲ p. 125

Syringe drivers ➲ p. 1030

Table 8.2 Quick conversions of oral morphine

From	To	Conversion	Example
Oral morphine (total dose) e.g. 10mg morphine 4 hourly = 60mg oral morphine in 24h	sc diamorphine	÷ by 3	**60** ÷ 3 = 20mg diamorphine by syringe driver over 24h
	sc morphine	÷ by 2	**60** ÷ 2 = 30mg morphine by syringe driver over 24h
	po oxycodone	÷ by 2	**60** ÷ 2 = 30mg oral oxycodone in divided doses over 24h

❗ If total 24h dose is equivalent to 120mg morphine or more—get specialist advice.

Opioid toxicity Intentional or unintentional overdose produces:

- Drowsiness or coma
- Pinpoint pupils
- Vomiting
- Hypotension
- Confusion—including auditory and/or visual hallucinations
- Respiratory depression:
  - If respiratory rate ≥ 8/min + the patient is easily rousable and not cyanosed—adopt a policy of 'wait and see'; consider ↓ or omitting the next regular dose of opioid. Stop syringe drivers temporarily to allow plasma levels to ↓, then restart at lower dose
  - If respiratory rate <8/min, and/or the patient is barely rousable/unconscious and/or cyanosed—dilute naloxone 400mcg to 10mL with sodium chloride 0.9%. Administer 0.5–1mL IV every minute until respiratory status is satisfactory. If respiratory function still does not improve, question diagnosis. Further doses may be needed later as naloxone is shorter acting than morphine
- Muscle rigidity/myoclonus—consider renal failure (can produce myoclonus alone). Treat with rehydration. Consider stopping other medication which may exacerbate myoclonus, switching opioid, or treating with clonazepam 2–4mg/24h depending on circumstances

Subacute overdosage Slowly progressive somnolence and respiratory depression—common in patients with renal failure. Withhold morphine for 1–2 doses then reintroduce at 25% lower dose.

Opioid toxicity may be ↑ by

- Renal failure
- Other change in disease status e.g. hepatic function, weight loss
- Dehydration
- Other analgesics e.g. NSAIDs
- Co-administration of amitriptyline

Management of drugs misuse ➔ p. 166

Further information

BNF 🖱 https://bnf.nice.org.uk/

Faculty of Pain Medicine Opioids Aware. 🖱 www.rcoa.ac.uk/faculty-of-pain-medicine/opioids-aware

Livewell with Pain 🖱 https://livewellwithpain.co.uk/

Online converter for fentanyl patches 🖱 www.globalrph.com/fentconv.htm

Neuropathic pain

Neuropathic pain occurs as a result of damage to neural tissue. Examples include post-herpetic neuralgia, complex regional pain syndrome (reflex sympathetic dystrophy), peripheral neuropathy (e.g. due to DM), compression neuropathy, and phantom limb pain.

Pain typically occurs in association with altered sensation, e.g. burning, stabbing, or numbness. Pain may also be provoked by non-noxious stimuli (allodynia), e.g. gentle heat or cold.

Diabetic neuropathy ➔ p. 330

Post-herpetic neuralgia Persistent neuropathic pain in a dermatome affected by shingles (➔ p. 628). May be severe.

Treatment of neuropathic pain

Trigeminal neuralgia[N] (➔ p. 531). The drug of choice for treatment of trigeminal neuralgia pain is carbamazepine (unlicensed indication). Often poorly tolerated. Oxcarbazepine is an alternative. If no response in <8wk, refer early for specialist assessment and advice on pain control.

Other neuropathic pain[N] Where possible, treat the underlying cause of the pain. If that is not possible, consider treatment with one of:

- Amitriptyline
- Duloxetine
- Gabapentin
- Pregabalin

For all these drugs, start at low dose and titrate the dose up according to response. Consider capsaicin cream if localized pain and the patient wishes to avoid, or cannot tolerate oral medication.

! Consider tramadol only if an acute rescue therapy is needed.

Review

After starting/changing medication Perform an early review after 1–2wk to check dosage titration, tolerability, and adverse effects.

Once established on medication Review regularly every 4–8wk to assess and monitor effectiveness of treatment. Ask about:

- Overall perception of improvement
- ↓ in pain (pain diaries may help)
- Adverse effects
- Sleep
- Ability to do everyday activities, e.g. work, driving
- Mood—depression/anxiety screening questionnaires may be helpful

If improvement is sustained over ≥6mo, consider gradual ↓ dose of medication over time.

Refer To a specialist pain clinic or other appropriate specialist service if:

- Severe pain or pain significantly limits activities
- Underlying health condition has deteriorated
- Inadequate response to first-line (trigeminal neuralgia) or second/third-line medication (all other neuropathic pain)

Antidepressants

Amitriptyline Proven treatment for neuropathic pain (unlicensed indication). Start at a dose of 25mg at 5–7 p.m.—10mg if elderly. ↑ dose by 10–25mg every 5–7d to a maximum of 75mg in a single dose as needed (higher doses under specialist supervision). Some patients do not derive benefit for 4–6wk.

Alternatives to amitriptyline include nortriptyline and imipramine. Both are given at an initial dose of 10–25mg in the evening; dose can be titrated up to 75mg as needed. May have fewer side effects than amitriptyline.

Duloxetine Effective treatment for neuropathic pain. Currently only licensed for the treatment of diabetic neuropathy. Usually less sedating than amitriptyline. Start with 60mg once daily, increasing to 60mg bd after 2–3wk if inadequate response.

Anticonvulsants

Carbamazepine Only recommended for treatment of trigeminal neuralgia. Start with 100–200mg 1–2×/d (less if elderly or frail). Build up dose slowly to the usual dose of 0.8–1.2 g daily in divided doses. Often poorly tolerated.

Pregabalin Licensed for treatment of neuropathic pain. Initially 150 mg/d in 2 divided doses, ↑ if necessary after 3–7d to 150mg bd, and again if needed after a further 7d to 300mg bd.

Gabapentin Effective for neuropathic pain but less clinically and cost-effective than pregabalin. Start with 300mg tds, increasing in steps of 300mg every 2–3d as needed to a maximum of 3.6g/d. Slower titration is advisable for the elderly or frail.

❗ Both gabapentin and pregabalin are drugs of abuse; monitor frequency of repeat prescriptions; be careful when issuing to temporary patients.

NSAIDs Sometimes effective for neuropathic pain—either because there is mixed nociceptive pain or because they ↓ inflammatory sensitization of nerves. There is considerable variation in individual patient tolerance and response (➔ p. 184).

Opioids Neuropathic pain often responds only partially to opioid analgesics. Of the opioids, oxycodone, tramadol and methadone are probably the most effective. Do not start regular treatment with opioids for neuropathic pain in primary care.

Capsaicin Active ingredient in chilli peppers. Licensed for treatment of neuropathic pain and applied locally as a cream 3–4×/d. Advise patients to apply the cream sparingly in a well-ventilated room and wash their hands after use. Side effects include intense burning at the site of application, and more rarely eye symptoms or sneezing. The cream should not be applied after a hot shower/bath as this intensifies the burning sensation.

Topical lidocaine Plasters impregnated with lidocaine 5% (Versatis®). Licensed for post-herpetic neuralgia. Apply daily for up to 12h, followed by a 12h plaster-free period; discontinue if no response after 4wk. Up to 3 plasters may be used to cover large areas; plasters may be cut.

Further information

BNF 🖱 https://bnf.nice.org.uk/
NICE (2013, updated 2018) Neuropathic pain. 🖱 www.nice.org.uk/guidance/cg173

Normal ageing

Of the UK's 65.6 million people, 18% are aged ≥65y, and 2.4% ≥80y. Life expectancy is ↑. Over the past 30y, the population aged >65y has grown by 23% with largest percentage growth among those >85y.

What is ageing? Ageing is a gradual series of changes over time that lead to loss of function of organs and cells, with the eventual outcome of death. Normal changes of ageing (Table 8.3). Individuals vary greatly in the rate at which they age. Several factors seem to influence this:

- Genetic makeup
- Psychological health
- Lifestyle—diet, physical exercise, smoking
- Socioeconomic factors
- Environment

Difficulties assessing the elderly

- Communication problems—hearing, cognition, speech
- Multiplicity of cause—1 symptom may be caused by different, concurrent processes, e.g. breathlessness as a result of COPD + heart failure
- Non-specific symptoms/signs—confusion, falls, or 'off legs' may be the only overt sign of underlying disease, e.g. UTI, MI, stroke
- Symptoms may be absent despite disease, and signs harder to elicit
- Polypharmacy (➔ p. 194) may result in side effects and interactions
- Laboratory tests may be unreliable—especially white cell counts and ESR (always check CRP)

Disease The ageing process is compounded by overt disease. This may affect functional capacity, quality of life, and independence, cause frailty, ↓ well-being and independence, and result in ↑ care and mobility needs.

Multiple morbidity ➔ p. 174. Older people are more likely to have several ongoing chronic illnesses that can act in combination to cause disability greater than either illness alone and/or result in:

- Direction of care at some problems with relative neglect of others
- Polypharmacy—➔ p. 194
- Involvement of multiple specialist teams which can cause inconvenience to the patient and family, and result in conflicting advice, and opposing opinions on cause/effect of symptoms

Frailty Elderly people are described as 'frail', as term used to describe individuals who are physically weak and fragile. Frailty can occur on a background of natural ageing or be precipitated by disease. It is not a disease/disability, but an inability to withstand physical/psychological stressors. Common features include:

- Feeling of exhaustion
- Unintentional weight loss (>5kg in a year)
- Weakness—measured by grip strength
- Low levels of physical activity
- Slow walking speed

Detecting frailty can enable support (➔ p. 198) to be put in place earlier, to enable frail people to remain well at home, and avoid crisis situations.

Table 8.3 Normal changes of ageing

System	**Clinical/functional effects**
Cardiovascular	Cardiac enlargement/left ventricular hypertrophy ↓ cardiac output → ↓ exercise capacity ↓ response of heart rate to exercise Systolic hypertension Left ventricular failure
Respiratory	↓ FEV_1/FVC and ↑ residual volume ↑ susceptibility to infection ↑ susceptibility to aspiration
Endocrine	↓ insulin sensitivity → impaired glucose regulation ↓ thyroid hormone production
Gastrointestinal	↑ in gastric acid production Constipation
Genitourinary	↓ glomerular filtration rate not reflected by ↑ creatinine Benign enlargement of the prostate (25–50% of men >65y) → prostatism (♂) Slowing of sexual function (♂ and ♀); erectile dysfunction (♂) Dry vagina and ↑ susceptibility to urinary infections (♀)
Musculoskeletal	Sarcopenia—↓ muscle strength/power, ↓ lean body mass (30–40%), ↑ fat body mass ↓ mobility; ↑ likelihood of falls ↑ osteoporosis/susceptibility to fractures
Nervous	Slower thought processes/reaction times General decline in performance ❗ Dementia is not a normal change of ageing
Vision	Presbyopia (difficulty focusing on near objects); ↓ visual acuity; cataract; impaired dark adaptation
Hearing	High-frequency hearing loss/presbycusis—deafness affects 80% of 80y-olds Degenerative changes in the inner ear leading to impairment of balance causing falls
Immune	Atrophy of the thymus Reduced immune function resulting in ↑ infectious disease, reactivation of latent disease (e.g. TB, shingles), ↑ cancer, and ↑ autoimmune disease
Skin/hair	Dry skin, wrinkles, tendency to bruise easily, and slower healing Greying of the hair ↓ sweating, heat generation, and heat conservation → heat stroke; hypothermia ↓ sensitivity to touch, pain, and temperature discrimination → burns and pressure sores

Further information

NHS England (2017) Toolkit for general practice in supporting older people living with frailty. 🔗 www.england.nhs.uk/publication/toolkit-for-general-practice-in-supporting-older-people-living-with-frailty

Falls in the elderly

Falls are a major cause of disability and the leading cause of mortality due to injury in people aged >75y. Tendency to fall ↑ with age. Assessment of a patient who has fallen is a common primary care emergency.

Risk factors for falls Recurrence ↑ with number of risk factors:

- ♀:♂≈2:1 in the over 75s
- ↑ age
- Multiple previous falls
- Disorders of gait or balance
- Visual impairment
- Cognitive impairment
- Low morale/depression
- High level of dependence
- ↓ mobility
- Foot problems
- Lower limb weakness or arthritis
- History of stroke or PD
- Use of psychotropic drugs, sedatives, diuretics or β-blockers
- Alcohol
- Environmental factors, e.g. loose rugs, poor lighting, ice, high winds
- Infection, e.g. pneumonia, UTI

Assessment Deal with the injuries first—ask about pain, loss of function, headache. Ask carers about behaviour. Check for bruising, ↓ function, confusion, BP, pulse, neurology, and fundi. Consider hypothermia if on the floor any duration.

Investigate the cause of the fall Consider:

- Physical problems Neurological problems (e.g. stroke); visual loss; cardiac abnormalities (e.g. arrhythmia, postural hypotension); muscular abnormalities (e.g. steroid-induced myopathy); skeletal problems (e.g. OA); infection (pneumonia, UTI)
- Environmental problems Climbing ladders to do routine maintenance; loose/holed carpets; slippery floor/bath; chair or bed too low

Management

- Treat any acute injury (20%). Exclude fracture (mainly Colles'/neck of femur). ❗ Subdural haematoma may take days/weeks to reveal itself
- Even if uninjured, older people might not be able to get up off the floor without help. The result may be a prolonged period of lying on the floor until help arrives. Apart from the indignity/helplessness this causes, 2° problems (e.g. pneumonia, pressure sores, hypothermia, UTI, and dehydration) may follow
- Perform/refer to a specialist falls service for a falls assessment
- Undertake measures to ↓ risk of falls or damage from falling

Further actions

- Refer to A&E—if significant head injury (➔ p. 1094); any suspicion of fracture; any other significant injury, e.g. lacerations
- Admit to the acute medical or elderly care team—if the cause of the fall was an acute medical problem, e.g. stroke
- Refer to the intermediate care (rapid response) team—if the patient is unable to cope at home or the patient/carer is worried about the possibility of further falls
- Refer to the specialist elderly care team or falls clinic—if the cause of recurrent falls remains unclear

Osteoporosis and prevention of fractures ➔ p. 482

Prevention of falls Falls are one of the biggest risk factors for fracture. All elderly people should have risk of falls assessed regularly. ❗ Any fall may seriously undermine an elderly person's confidence and cause worry about the possibility of recurrence. As a result, there may be restriction of activities → ↓ fitness and ↑ dependency on others.

Is a falls assessment needed? Ask if patients fall—they may not volunteer the information spontaneously.

The timed get-up-and-go test ❗ May use usual walking aid.

- Start with the patient sitting in a straight-backed chair of comfortable height with arms
- Ask the patient to rise from the chair, walk to a line 10 feet (3m) away, turn around, return to the chair, and sit down again
- Start timing while the patient is sitting; end timing when the patient has sat down again
- A time of ≥13sec predicts ↑ falls risk

Falls assessment If available, refer to a specialist falls service. *Record*:

- Frequency and history of circumstances around any previous falls
- Drug therapy: polypharmacy, hypnotics, sedatives, diuretics, antihypertensives may all cause falls
- Assessment of gait and balance, including abnormalities due to foot problems or arthritis, and motor disorders, e.g. stroke, PD
- Examination of basic neurological function, including vision, mental status (impaired cognition and depression), muscle strength, lower extremity peripheral nerves, proprioception, and reflexes
- Assessment of basic cardiovascular status including BP (exclude postural hypotension), heart rate, and rhythm
- Assessment of environmental risk factors, e.g. poor lighting particularly on the stairs, loose carpets or rugs, badly fitting footwear or clothing, lack of safety equipment such as grab rails, steep stairs, slippery floors, or inaccessible lights or windows

Measures to ↓ risk of falls and damage from falling

- Correct vision, if possible
- Correct postural hypotension—alter medication; consider compression stockings—but many elderly people cannot apply stockings tight enough to be of any use themselves
- Treat other medical conditions, e.g. refer to cardiology if arrhythmia
- Review medication and discontinue/alter inappropriate medication
- Remove environmental hazards—arrange bath at a day centre, refer to OT to identify/correct hazards in the home, e.g. remove loose carpets, wheeled trolley for use indoors, commode/urine bottle at night, etc.
- Liaise with other members of the PHCT and social services to provide additional support if needed; refer to local council for 'carephone' or alarm system to call for help if any further falls
- Refer to rehabilitation/physiotherapy to improve confidence after falls and for weight-bearing exercise (focusing on strength and flexibility) and balance training (↓ risk of falls). Use of hip protectors ↓ fracture risk in patients at high risk but compliance is a problem

Prescribing for older people

Use of medicines ↑ as people get older; 1 in 3 NHS prescriptions are for patients >65y and 90% of these prescriptions are for repeat medication. Adverse drug events are common reasons for hospital admission in the >75 age group; many are avoidable. Regular review is essential. Problems commonly encountered:

Polypharmacy Elderly people often have multiple problems. It is easy to keep adding drugs for each new problem resulting in polypharmacy. This ↑ confusion about drug regimens, and results in poor concordance and multiple interactions/side effects.

- Before prescribing a new drug consider whether it is necessary—avoid treating normal changes of ageing; use non-pharmacological therapies wherever possible; avoid 'a pill for every ill' approach and try to treat the underlying condition not the symptoms
- Balance the potential risks of the drug against the benefits. Drug trials of efficacy of medication often exclude older participants—the applicability of evidence to elderly patients cannot be assumed. For prophylactic medication (e.g. warfarin, statins), consider the likelihood of concordance and benefits in the context of the whole person (including other co-morbidities)
- Review medication regularly. Stop ineffective/redundant drugs and consider if the overall drug regimen can be simplified

Form of the medicine Swallowing tablets can be difficult for elderly people. Consider using liquid preparations/giving explicit advice to take medication with plenty of water and sitting upright.

Confusion after discharge Up to ½ all patients are inadvertently prescribed the wrong medication after hospital discharge.

Drug hoarding/self-medication Especially if recent changes in medication it is common for elderly people to have a back stock of drugs and continue taking their old drugs alongside new ones. A written list may be helpful. Many elderly people also self-medicate extensively with OTC preparations. If necessary, do a home visit to sort out the drugs.

↑ susceptibility to side effects Common due to altered:

- Pharmacodynamics ↑ susceptibility to GI side effects (e.g. constipation with opioids; gastric irritation with NSAIDs) and ↑ sensitivity to effects of CNS drugs, e.g. benzodiazepines, opioids—use with care
- Pharmacokinetics ↓ renal function is particularly important—always assume any elderly person has moderate renal impairment

Social and personal factors Low level of home support; physical factors, e.g. poor vision, poor hearing, or poor manual dexterity; and mental state, e.g. confusion/disorientation, depression—can all affect ability of an older person to take medication.

Specific medicines The Beer's list is a list of agents to be avoided/used with extreme caution in elderly patients. It can be accessed via 🖱 www.dcri.duke.edu/ccge/curtis/beers.html

Guidelines for prescribing for the elderly

Think before prescribing

- Is the drug needed?
- Is there another non-pharmacological way of managing the problem?
- Are you treating the underlying condition or the symptoms of it?
- What are the pros and cons of the patient taking this drug?
- What is the evidence base for its use in this age group?
- Will the patient be able to take the drug (formulation; packaging)?
- Will the patient be concordant?
- Will the patient comply with any necessary monitoring?

Limit the range of drugs you use

Prescribe from a limited array of drugs that you know well.

Repeats and disposal

- Tell patients how to get more tablets and monitor frequency of repeat prescriptions; review repeat prescriptions regularly (➔ p. 118)
- Tell patients what to do with any left-overs if a drug is stopped

↓ *the dose*

- Start with 50% of the adult dose
- Avoid drugs likely to cause problems e.g. hypnotics

Review regularly

- Consider on each occasion whether each drug could be stopped or the regimen simplified
- Consider lowering dosage of drugs if renal function is deteriorating
- Involve carers, community pharmacists, and other PHCT members

Simplify regimens

Use od or bd regimens wherever possible; avoid polypharmacy.

Explain clearly

- Decision aids can be helpful, e.g. 🖱 www.anticoagulation-dst.co.uk/
- Put precise instructions on the drug bottle—avoid 'use as directed'
- Give written instructions about how the drug should be taken
- Ensure explanations are given to carers as well, if appropriate

Consider method of administration

- Bottles with child-proof tops are often impossible for arthritic hands to open. Suggest the patient asks the chemist for a standard screw cap
- Drug administration boxes in which the correct tablets are stored in slots marked with the day and time of administration can be helpful. Available from pharmacists and can be filled by the patient, a carer, friend, or relative, or the pharmacist
- Medication reminder charts can also help

Further information

Gallagher P, et al. (2008) STOPP (Screening Tool of Older Persons Prescriptions) and START (Screening Tool to Alert doctors to Right Treatment). Consensus validation. *Int J Clin Pharmacol Ther* **46**:72–83.
Lavan AH, et al. (2017) STOPPFrail (Screening Tool of Older Persons Prescriptions in Frail adults with limited life expectancy): consensus validation. *Age Ageing* 1;**46**:600–7.

Elderly care and disability management

> '*Use strengthens, disuse debilitates*'
>
> Hippocrates (460–357 BC)

13–14% of the population has some disability. This is ↑ as populations age and people survive longer with disability. Many more are just elderly and frail. 35% of people aged >80y cannot live an independent life. Most patients are best managed by a MDT in their home environment (if practicable) with a problem-oriented approach. Good interdisciplinary communication and coordination is essential and many patients benefit from specialist rehabilitation services. Psychological and sociocultural aspects are as important as medical aspects of care.

Role of the GP within the MDT Maintain an open door policy and encourage patients and carers to seek help for problems early. Try to become familiar with patients' diseases, even if rare. Information alone can improve outcome.

Consider
- Can physical symptoms be improved?
- Can psychological symptoms be improved (including self-esteem)?
- Can functioning within the home be improved (aids and adaptations within the home, extra help)?
- Can functioning in the community be improved (mobility outside the home, work, social activities)?
- Can the patient's or carer's financial state be improved?
- Does the carer need more support?

❗ If progress is slower than expected, or stalls, consider other medical problems (e.g. anaemia, hypothyroidism, dementia), a neurological event, depression, or communication problems (e.g. poor vision/hearing).

Principles of care

- **Use of assessments/measures** Central to the management of disability. Use validated measures accepted by all team members (e.g. Barthel index; PHQ-9—➔ p. 979). Reassess regularly
- **Multidisciplinary teamwork** Good outcomes are associated with clinicians and other involved health and social care professionals working as a team towards a common goal with patients and their families (or carers) included as team members
- **Goal-setting** Goals must be meaningful and challenging—but achievable. Use short- and long-term goals. Involve the patient ± carer(s). Regularly renew, review, and adapt
- **Underlying approach to therapy** All approaches focus on modification of impairment and improvement in function in everyday activities. May include drugs, physical or psychological therapies, ± social support
- **Referral for specialist support** Consider specialist medical or rehabilitation services; social services (if not already part of the MDT); voluntary organizations and self-help groups; Citizen's Advice

Maintaining independence ➔ p. 198 **Carers** ➔ p. 200

Common neurological rehabilitation problems ➔ p. 554

Learning disability health checks People with a learning disability have poorer physical/mental health and die younger. Annual health checks can be offered as a Directed Enhanced Service to patients aged >14y on the practice learning disability register. Standard templates are available within practice software systems. Health checks aim to:

- Ensure all available health prevention measures are offered, e.g. screening, routine vaccinations, contraception
- Identify health conditions that might otherwise go undetected
- Optimize management of ongoing conditions
- Identify patients/carers who may need more support

Adult safeguarding Means to protect a person's right to live in safety, free from abuse, harm, and neglect. Includes proactive and reactive measures and recognizes that adults may choose to make unwise decisions. An adult at risk is any person who is ≥18y and at risk of abuse, harm, or neglect because of their needs for care and/or support.

Types of abuse Include:

- Physical e.g. hitting, misuse of medication or restraint
- Sexual e.g. rape, sexual acts to which the adult has not consented
- Psychological e.g. threats of harm/abandonment, humiliation, blaming
- Financial e.g. theft, exploitation, fraud
- Neglect e.g. ignoring health/care needs, inadequate nutrition

Six safeguarding principles Underpin adult safeguarding:

- Empowerment
- Prevention
- Proportionality
- Protection
- Partnership
- Accountability

The role of the GP GPs must undergo regular adult safeguarding training. This is assessed as part of their annual appraisal. They must:

- Be able to recognize potential signs of abuse
- Work as part of the MDT (including local adult safeguarding services) to take all necessary action to safeguard their patients
- Reflect on safeguarding events and take any necessary actions to improve adult safeguarding within their own organizations

Deprivation of Liberty Safeguards (DoLS) ➔ p. 1106

Elder abuse ➔ p. 87 **Domestic violence** ➔ p. 86

Modern slavery ➔ p. 88 **Female genital mutilation** ➔ p. 710

'Prevent' strategy ➔ p. 89

Organizational abuse Usually affects people in residential care settings (e.g. hospitals, care homes) but can occur in relation to paid care in people's own homes. Individuals' wishes and needs are sacrificed for the smooth running of the service/organization, e.g. no choice of food, shared clothing, forced early bed times. If suspected, raise with the service provider in the first instance (with the patient's/carer's permission if possible). If no action is taken, refer to local social services adult safeguarding lead.

Further information

RCGP Safeguarding adults at risk of harm toolkit. 🖱 www.rcgp.org.uk/clinical-and-research/resources/toolkits/safeguarding-adults-at-risk-of-harm-toolkit.aspx

Maintaining independence

Maintaining independence is important. It gives individuals autonomy, a sense of purpose and achievement, and allows them to make their own choices. Lack of independence can result in a feeling of 'being a burden', boredom, frustration, loneliness, and social isolation.

Equipment and adaptations If difficulty coping at home due to disability or age, anyone can request a needs assessment by an occupational therapist via their local social services department. This enables provision of equipment/adaptations necessary to maintain independence.

Benefits ➔ p. 104

Help to use the telephone Many people have difficulty using the telephone either because of physical impairments or communication problems. British Telecom can provide suggestions about how to address many of these issues—🖱 www.bt.com/includingyou/index.html

Alarm systems Enable anyone who is alone at times to call for help, even when they cannot reach a telephone. Suitable for anyone capable of using an alarm system. Arrange via local social services or housing departments. Alternatively charities for the elderly have schemes (e.g. Age UK Personal Alarm ☎ 0800 030 4385).

Walking aids Wide variety—from simple walking sticks and crutches, to a range of wheeled and unwheeled walking frames. Physiotherapists can advise on the best mobility aid for each individual patient. Can be obtained on loan from the NHS or purchased by the individual.

Wheelchairs If needed short term, available on loan from local Red Cross branches. The NHS can provide a wheelchair for anyone requiring one for >3mo. Referral must be made by an authorized healthcare practitioner (e.g. GP) to the local NHS Wheelchair Service. After assessment individuals can opt to have an NHS wheelchair or be provided with a voucher to put towards the cost of their own. *Directory of NHS wheelchair service centres*: 🖱 www.wheelchairmanagers.nhs.uk

Community transport services Available in many areas for people who have difficulty using public transport. Include door-to-door transport to attend appointments, trips to shopping centres, and other outings. Local arrangements vary.

Free bus passes ❗ May be restrictions on time of travel.

Disabled people are entitled to free bus passes in England through the English National Concessionary Travel Scheme if they meet eligibility criteria (Box 8.1). Similar schemes operate in Scotland, Wales, and Northern Ireland. A carer may also be issued with a free pass if the individual cannot travel alone. Applications are made via local authorities, but the bus pass can be used anywhere within the country it was issued. A GP letter to confirm eligibility may be required.

Elderly people can also apply for free bus passes from 60y in London (also valid for Underground), Scotland, Northern Ireland, and Wales, and from the women's state retirement age elsewhere in England.

Disability and driving ➔ p. 99

Box 8.1 Criteria for application for a free bus pass in England

A disabled person is likely to get a concessionary bus pass if he or she:

- Is blind or partially sighted
- Is deaf or unable to speak
- Is unable to walk very far because of a disability, illness, or injury
- Is unable to use his/her arms (or does not have arms)
- Has severe learning disability
- Has been refused a driving licence for a health reason (but not because of problems with drugs or alcohol)

Blue Badge Scheme Entitles the holder to park:

- In specified disabled spaces
- Free of charge or time limit at parking meters or other public places where waiting is limited
- On single yellow lines for up to 3h (no time limit in Scotland)

The disabled person does not have to be the driver but the badge should not be used if the disabled person is not in the car. Applications are made via the local authority. Automatically eligible if >2y old and one of the following criteria apply:

- Registered as blind
- Receiving higher rate mobility component of DLA, or receiving PIP and scored ≥8 points in the 'moving around' area of the PIP assessment
- Getting War Pensioners' Mobility Supplement or received a lump sum payment as part of the Armed Forces Compensation Scheme (tariffs 1–8), and certified as having a permanent and substantial disability

People with permanent problems walking, or who cannot use their arms may be eligible and should apply—each application is judged on its merits.

Motability (🖱 www.motability.co.uk) Qualifying benefits (must have ≥12mo award length remaining):

- Higher rate mobility component of DLA
- Enhanced rate mobility component of PIP
- War Pensioners' Mobility Supplement, or
- Armed Forces Independence Payment

Mobility payments associated with state benefits can be used to lease a car, wheelchair-accessible vehicle, scooter, or powered wheelchair. Includes many special adaptations, insurance, breakdown assistance, servicing, and maintenance. Grants may also be available for additional adaptations or driving lessons. ❗ The driver does not have to be the claimant.

Road Tax Exemption Should be received automatically by anyone receiving any of the eligibility benefits listed for the Motability scheme.

Patient information and support

Age UK Wide range of information and factsheets. ☎ 0800 055 6112 🖱 www.ageuk.org.uk

Citizens Advice 🖱 www.adviceguide.org.uk

Disabled Living Foundation Advice about equipment and appliances. ☎ 0300 999 0004 🖱 www.dlf.org.uk

Scope. Disability information line. ☎ 0808 800 3333 🖱 www.scope.org.uk

Carers

Who is a carer? A carer is someone of any age who provides unpaid support to family or friends who could not manage without this help. This could be caring for a relative, partner, or friend who is ill, frail, disabled, or has mental health or substance misuse problems. Anyone can become a carer. ~2 million people/y move in/out of caring roles.

Young carers Children and young people who assume inappropriate responsibilities to look after someone who has an illness, a disability, or is affected by mental ill health or substance misuse. Young carers often take on practical and/or emotional caring responsibilities that would normally be expected of an adult.

> ❗ Some carers do not regard themselves as carers, or may dislike the label 'carer', believing that it can detract from their identity as a parent, child, partner, or sibling to the person that they care for. It is also important not to confuse carers with paid care workers.

How many carers are there? 12% of adults in the UK are carers. 1.2 million provide care for >50h/wk; those aged >65y account for 1 in 3 of those providing >50h care each week and many have their own health problems too. There are also around 1 million young carers.

Carers as partners in care Carers know the people that they care for better than anyone else. Involving carers is important in order to identify problems that may require intervention; plan patient care; and improve concordance with care plans.

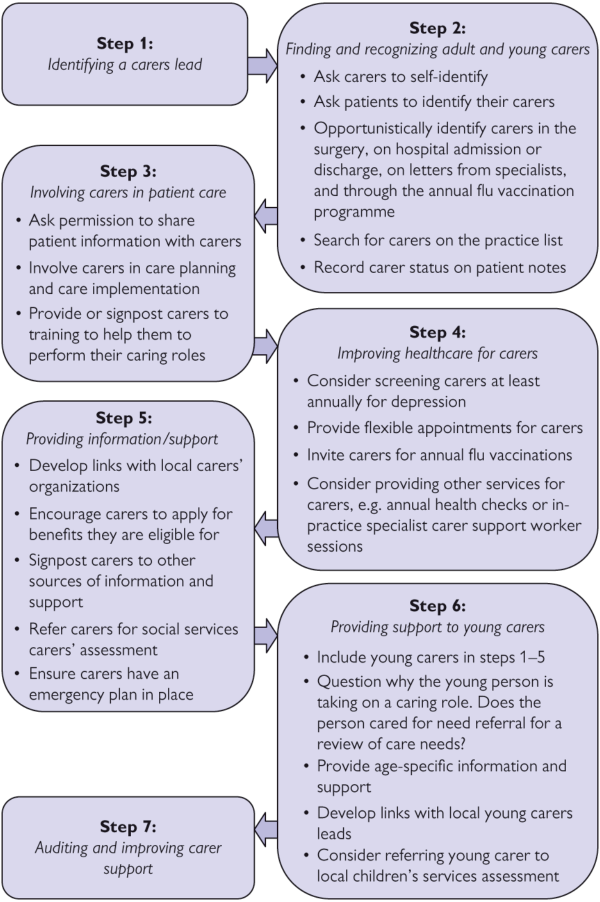
What problems do carers have as a result of their roles? Many carers gain great personal satisfaction from their caring role and want to continue caring, but they suffer adverse consequences too:

- Psychological ↑ stress and depression/anxiety; abuse from the person being cared for; young carers have ↑ risk of bullying/self-harm
- Physical health ↑ mortality; ↑ morbidity from CVD; ↑ risk of back and other musculoskeletal injury
- Social activity restriction Deterioration of relationships with other family members, social isolation
- Employment/schooling ↓ ability to work, ↓ promotion prospects, poor performance, and ↑ absenteeism at school
- Financial The more care provided, the more likely a carer is to be in financial difficulty; 55% are in debt but <50% claim all the benefits they are eligible for

Supporting carers in general practice See Figure 8.6

Benefits For sickness/disability/carers ➔ p. 108; low income ➔ p. 104

Social services assessment Every carer has a right to ask for a full assessment of their needs by the social services. Emergency planning to provide substitute care in the event of a crisis is part of that assessment.



Step 1:
Identifying a carers lead

Step 2:
Finding and recognizing adult and young carers

- Ask carers to self-identify
- Ask patients to identify their carers
- Opportunistically identify carers in the surgery, on hospital admission or discharge, on letters from specialists, and through the annual flu vaccination programme
- Search for carers on the practice list
- Record carer status on patient notes

Step 3:
Involving carers in patient care

- Ask permission to share patient information with carers
- Involve carers in care planning and care implementation
- Provide or signpost carers to training to help them to perform their caring roles

Step 4:
Improving healthcare for carers

- Consider screening carers at least annually for depression
- Provide flexible appointments for carers
- Invite carers for annual flu vaccinations
- Consider providing other services for carers, e.g. annual health checks or in-practice specialist carer support worker sessions

Step 5:
Providing information/support

- Develop links with local carers' organizations
- Encourage carers to apply for benefits they are eligible for
- Signpost carers to other sources of information and support
- Refer carers for social services carers' assessment
- Ensure carers have an emergency plan in place

Step 6:
Providing support to young carers

- Include young carers in steps 1–5
- Question why the young person is taking on a caring role. Does the person cared for need referral for a review of care needs?
- Provide age-specific information and support
- Develop links with local young carers leads
- Consider referring young carer to local children's services assessment

Step 7:
Auditing and improving carer support

Figure 8.6 Practice action plan for supporting carers

Further information

RCGP (2011) Supporting carers: an action guide for GP and their teams. 🖱 www.rcgp.org.uk

Carer support

Carers Trust 🖱 www.carers.org
Carers UK 🖱 www.carersuk.org

Chapter 9

Cardiology and vascular disease

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Symptoms and signs of CVD

Chest pain ➔ p. 1064
Blood pressure ➔ p. 218
Breathlessness or dyspnoea ➔ p. 264
Crackles in the chest ➔ p. 269

Peripheral oedema Swelling of the ankles/legs (or sacrum if bed bound) occurs when the rate of capillary filtration > rate drainage.

- ↑ capillary filtration occurs due to ↑ venous pressure, hypoalbuminaemia, or local inflammation
- ↓ drainage occurs due to lymphatic obstruction

Consider whether swelling is acute or chronic, symmetrical or asymmetrical, localized or generalized. Ask about associated symptoms, e.g. breathlessness. Treat according to cause. *Causes*:

Acute

- DVT
- Superficial thrombophlebitis
- Joint effusion/haemarthrosis
- Arthritis
- Cellulitis
- Haematoma
- Baker's cyst
- Fracture
- Acute arterial ischaemia
- Dermatitis

Chronic

- Gravitational oedema, e.g. due to immobility—common in the elderly. Advise elevation of feet above waist level, support stockings (ideally apply stockings before getting out of bed), avoid standing still. Diuretics are not a long-term solution
- Heart failure
- Hypoproteinaemia, e.g. nephrotic syndrome
- Idiopathic oedema
- Reflex sympathetic dystrophy
- Post-thrombotic syndrome
- Chronic venous insufficiency/venous obstruction
- Lipodermatosclerosis
- Lymphoedema—infection, tumour, trauma
- Congenital vascular abnormalities

Pulmonary oedema Accumulation of fluid in the pulmonary tissues and air spaces. *Causes include*:

Cardiac/vascular

- Left heart failure
- Mitral stenosis
- MI
- Hypertension
- Pulmonary venous obstruction
- IV fluid overload

Other

- High altitude
- Kidney failure
- Nephrotic syndrome
- Cirrhosis
- Lymphatic obstruction, e.g. due to tumour

Lung

- Pneumonia
- PE
- Pneumonitis due to inhalation of toxic substances, e.g. gases, radiation

Cyanosis Dusky blue skin.

Central cyanosis Cyanosis of mucus membranes, e.g. mouth. *Causes*:

- Lung disease resulting in inadequate oxygen transfer (e.g. COPD, PE, pleural effusion, severe chest infection)

- Shunting from pulmonary to systemic circulation (e.g. Fallot's tetralogy, PDA, transposition of the great arteries)
- Inadequate oxygen uptake (e.g. met- or sulfhaemoglobinaemia)

Peripheral cyanosis e.g. cyanosis of fingers. *Causes*: as for central cyanosis plus:

- Physiological (cold, hypovolaemia)
- Local arterial disease (e.g. Raynaud's syndrome)

❗ Feet can be a dusky blue colour due to venous disease. If this occurs without central cyanosis it does not imply abnormal oxygen saturation.

Mitral facies Dusky bluish red flushing of the cheeks (a form of peripheral cyanosis) associated with a low cardiac output.

Clubbing Loss of the angle between nail fold and plate, bulbous fingertip, and the nail fold feels boggy—➔ p. 577.

⚠ Refer any patient with unexplained nail clubbing for urgent CXR[N].

Jugular venous pressure Observe internal jugular vein at 45° with head turned slightly to the left. Vertical height is measured in relation to the sternal angle. Raised if >4cm. Causes of ↑ JVP:

- Fluid overload
- Right heart failure and CCF
- SVC obstruction (non-pulsatile)
- Tricuspid or pulmonary valve disease
- Pulmonary hypertension
- Arrhythmia—AF or atrial flutter, complete heart block
- ↑ intrathoracic pressure, e.g. pneumothorax, PE, emphysema

Kussmaul's sign The JVP usually drops on inspiration along with intrathoracic pressure. The reverse pattern is called Kussmaul's sign. Caused by raised intrathoracic pressure or constrictive pericarditis.

Signs of infective endocarditis

- **Infective** Fever, weight ↓, clubbing, splenomegaly, anaemia
- **Cardiac** Murmurs (particularly new murmurs) ± heart failure
- **Embolic** Neurologic deficit due to stroke
- **Vasculitic** Microscopic haematuria, splinter haemorrhages, conjunctival haemorrhages, Roth's spots (retinal vasculitis), Osler's nodes (painful lesions on finger pulps), Janeway lesions (palmar macules)

Signs of hypercholesterolaemia

Corneal arcus Whitish opaque line surrounding the margin of the cornea, separated from it by an area of clear cornea. Rarely congenital—more commonly occurs bilaterally in patients >50y (*arcus senilis*). Sometimes associated with ↑ blood lipids—particularly familial hypercholesterolaemias. Check lipids. If lipids are normal, no treatment is needed.

Xanthomata Localized collections of lipid-laden cells. Appear as yellowish coloured lumps. Often caused by ↑ lipids. Commonly seen on the eyelids (*xanthelasma*), on the skin, or in tendons (appear as mobile nodules in the tendon).

Examining the heart

Apex beat Normal position is in the 5th intercostal space, in the midclavicular line. Moved sideways/inferiorly if the heart is enlarged (e.g. CCF) or displaced (e.g. pneumothorax). May not be palpable if the patient is obese, has hyperexpanded lungs (e.g. COPD) or a pericardial effusion. In infants/children apex beat is superior/more lateral.

Parasternal heave Detect by placing the heel of the hand over the left parasternal region. If present, the heel of the hand is lifted off the chest wall with each heartbeat. *Causes*: Usually right ventricular enlargement—rarely left atrial enlargement.

Heart sounds Table 9.2. Low/medium-frequency sounds (e.g. 3rd/4th heart sounds) are more easily heard with the bell applied lightly to the skin. High-frequency sounds (e.g. 1st/2nd heart sounds and opening snaps) are more easily heard with a diaphragm.

Heart murmurs Due to abnormalities of flow within the heart and great vessels. Very common. Often incidental findings. Described by:

- **Location** Where heard loudest
- **Quality** e.g. blowing, harsh
- **Intensity** Graded out of 6 (1—virtually undetectable; 6—heard by an observer with no stethoscope). Grades 4–6 are usually palpable (*thrills*)
- **Timing** Systolic or diastolic
- **Radiation** Does the murmur spread elsewhere, e.g. to axilla, carotids

⚠ *Red flag symptoms*

- Cyanosis
- Breathlessness
- Lethargy/tiredness
- Collapse
- Weight loss (or failure to thrive)

Always refer for Echo. Differential diagnosis—Table 9.1.

Table 9.1 Differential diagnosis of heart murmurs

Type of murmur	Description	Causes
Ejection systolic murmur	↑ to reach a peak midway between the heart sounds.	• Flow murmurs, e.g. children, pregnancy, with fever, during/after exercise • Aortic stenosis or sclerosis (➔ p. 251) • Pulmonary stenosis (➔ p. 251) • HOCM (➔ p. 248)
Pan-systolic murmur	Uniform intensity between the 2 heart sounds. Merges with 2nd heart sound	• Mitral valve regurgitation/prolapse (➔ p. 250) • Tricuspid regurgitation (➔ p. 251) • VSD (➔ p. 252) • ASD (➔ p. 252)
Early diastolic murmur	Occurs just after the 2nd heart sound. High pitched. Easily missed.	• Aortic regurgitation (➔ p. 251) • Pulmonary regurgitation (➔ p. 251) • Tricuspid stenosis (mitral stenosis coexists)
Mid-diastolic murmur	Midway between 2nd heart sound of 1 beat and 1st of the next. Rumbling/low pitch	• Mitral stenosis (➔ p. 250) • Aortic regurgitation. (Austin Flint murmur—➔ p. 251)

Table 9.2 Heart sounds, abnormalities and their causes

Heart sound		Causes
1st heart sound Heard loudest at the apex. Caused by closing of the mitral and tricuspid valves	Soft	Mitral regurgitation, low BP, rheumatic carditis, severe heart failure, LBBB
	Loud	AF, tachycardia, atrial premature beat, mitral stenosis
	Variable intensity	Varying duration of diastole, complete AV block
	Split	RBBB, paced beat from the left ventricle, left ventricular ectopics, ASD, Ebstein's anomaly, tricuspid stenosis
2nd heart sound Caused by closure of the aortic (A2) and pulmonary (P2) valves A2 and P2 split on inspiration so that P2 is heard after A2	Soft	A2—calcification of the aortic valve, dilatation of the aortic root P2—pulmonary stenosis
	Loud	A2—↑ BP; thin patients P2—pulmonary hypertension, ASD
	Wide splitting	May be the result of early A2 or delayed P2 Early A2—mitral regurgitation; VSD Delayed P2—RBBB, pulmonary stenosis, ASD, right ventricular failure
	Reversed splitting	A2 is delayed. P2 occurs before A2 so the split between the sounds ↓ on inspiration Delayed A2—LBBB, systolic hypertension, HOCM, severe aortic stenosis, PDA, left heart failure
	Single	Calcification of the aortic valve, pulmonary stenosis, Fallot's tetralogy, Ebstein's anomaly, pericardial effusion, large VSD, obesity, emphysema
Clicks and snaps	Early systolic	Caused by opening of the aortic or pulmonary valves Aortic—aortic stenosis, bicuspid valve Pulmonary—pulmonary stenosis, pulmonary hypertension
	Mid/late systolic	Mitral valve prolapse
	Diastolic	Caused by opening of the mitral or tricuspid valves. Silent in the healthy heart Mitral—mitral stenosis, rapid mitral flow, e.g. PDA, VSD, severe mitral regurgitation Tricuspid (rare)—rheumatic stenosis, ASD
3rd heart sound Heard in diastole after the 2nd heart sound	Right ventricle	Loudest at lower left sternal edge. Never normal. Causes: right heart failure, tricuspid regurgitation, ASD, constrictive pericarditis
	Left ventricle	Loudest at the apex when inclined to the left. Can be normal in children and pregnancy. Other causes: LVF, mitral regurgitation, anterior MI
4th heart sound Heard in late diastole		Maximal at the apex or lower left sternal edge. Never normal. Causes: ventricular hypertrophy or fibrosis and HOCM

Examination of the arterial system

The main conditions affecting the abdominal and peripheral arteries are:

- Aneurysms (➔ p. 254)
- Atherosclerosis resulting in ischaemia of the legs and intermittent claudication, atrophic changes, and/or rest pain
- Embolization resulting in acute ischaemia of the limbs

General scheme

- Look at the limbs—are there any signs of ischaemia? Are the extremities warm or cold? What colour are they?
- Examine the abdomen looking for a pulsatile mass suggesting abdominal aortic aneurysm (➔ p. 254). Auscultation may reveal a bruit
- Check the peripheral pulses

⚠ Tenderness on palpation of an abdominal aortic aneurysm suggests need for urgent operative repair.

Blood pressure ➔ p. 216

Carotid pulse Ask the patient to lie supine with head/neck at 45° to the horizontal. When assessing the carotid pulse, consider:

Rate

- Tachycardia >100bpm—➔ p. 238
- Bradycardia <60bpm—➔ p. 242

Rhythm

- Irregularly irregular AF, multiple ectopics
- Regularly irregular 2nd-degree heart block

Character and volume Always assess with a central pulse, e.g. carotid or femoral.

- Small volume Shock, pericardial tamponade, aortic stenosis (slow-rising)
- Large volume Hyperdynamic circulation (e.g. pregnancy), aortic incompetence (water-hammer, collapsing pulse), PDA
- Pulsus paradoxus Pulse weakens in inspiration by >10mmHg—asthma, cardiac tamponade, pericarditis

Carotid bruits May signify stenosis (>30%) often near the origin of internal carotid. Heard best behind the angle of the jaw. Usual cause is atheroma.

Peripheral pulses

Location Table 9.3

Examination Check whether each pulse is present. If present check:

- Rate
- Rhythm
- Amplitude
- Compare pulses in the 2 legs/2 arms

Check for radiofemoral delay—palpate radial and femoral pulses simultaneously; delay suggests coarctation of the aorta.

Table 9.3 Location of the limb pulses

Pulse	Location
Brachial	~2cm medial to the central point of the antecubital fossa over the elbow skin crease.
Radial	~½–1cm on the radial (lateral) side of the flexor carpi radialis tendon at the wrist
Femoral	Below inguinal ligament; ⅓ of the way up from pubic tubercle
Popliteal	With knee flexed at right angles palpate deep in the midline
Posterior tibial	1cm behind medial malleolus
Dorsalis pedis	Variable—on the dorsum of the foot just lateral to the tendons to the big toe. ❗ Many healthy people have only 1 foot pulse

Check for bruits over the femoral and/or carotid pulses—these indicate disturbed blood flow—usually 2° to narrowing due to atherosclerosis.

⚠ Character and waveform of the pulse should *only* be assessed using the femoral or carotid pulse.

Signs of ischaemia

Acute ischaemia Acutely pale, cold, and pulseless limb—➔ p. 1108. Refer immediately—keep the limb cool in the interim.

Chronic ischaemic changes

- Atrophic skin changes—pallor, cool to the touch, hairless, shiny
- On lowering, the leg turns a dusky blue-red colour; on elevation, pallor and venous guttering
- Ulceration—check under the heel and between the toes
- Swelling suggests the patient is sleeping in a chair to avoid rest pain or, rarely, pain from deep infection
- Absent foot pulses—if pulses are present consider alternative diagnosis
- Ankle–brachial pressure index <0.95

Checking the ankle–brachial pressure index (ABPI)

- Check BP in one arm (➔ p. 216). The systolic measurement is the brachial pressure (B)
- Then inflate a BP cuff around the lower calf just above the ankle
- Using a Doppler ultrasound probe, record the maximum cuff pressure at which the probe can still record a pulse (ankle pressure—A)
- Calculate the ABPI by dividing the ankle pressure by the brachial pressure, i.e. ABPI = A ÷ B

Interpretation of ABPI results

- ABPI <0.8—ischaemia
- ABPI <0.5—critical ischaemia

❗ Arterial calcification (e.g. due to DM) can result in falsely elevated ankle pressure readings.

Cardiac investigations

Electrocardiogram (ECG) Graphic recording of electric potentials generated by the heart. Most surgeries now have ECG machines that interpret themselves and print out their findings. Analysis is easier but it is still important to be able to understand the significance of abnormalities and check computer analysis in the clinical context.

Interpreting ECGs Many mistakes in ECG interpretation are errors of omission so a systematic approach is best. *Check*:

- Standardization (calibration) and technical features (including lead placement and artefacts)
- Heart rate—usual speed (25mm/sec). Each big square represents 0.2sec; each small square, 0.04sec. Rate = 300 ÷ R–R interval in large squares
- Rhythm—regular/irregular
- PR interval—normal if <0.2sec
- QRS interval—abnormal if >0.12sec
- QT interval—varies with rate. At 60bpm normal if 0.35–0.43sec
- P waves—present or absent, shape
- QRS voltages—height of complexes—see Table 9.4, ➔ p. 212
- Mean QRS electrical axis—sum of all ventricular forces during ventricular depolarization. Normal axis: −30° to +120°
  - If more −ve = left axis deviation; if more +ve = right axis deviation
  - **Rule of thumb 1** If the majority of the QRS complex is above the baseline (+ve) in leads I and II the axis is normal
  - **Rule of thumb 2** The axis lies at 90° to a QRS complex where the height above the baseline = height below the baseline
- Precordial R-wave progression
- Abnormal Q waves—>25% of the succeeding R-wave and/or >0.04sec wide
- ST segments—elevation/depression, shape
- T waves—height, inversion, shape
- U waves—small, rounded deflection (≤1mm), follows T wave and usually has the same polarity

Brief guide to common ECG changes Table 9.4, ➔ p. 212

24h ambulatory ECG ECG monitoring equipment is worn for 24h. Continuous monitoring may detect intermittent arrhythmia or ischaemia.

Cardiac MRI/magnetic resonance angiography Used as the first -line investigation to assess patients with chest pain for suspected IHD. Increasingly used in 2° care to provide detailed structural information about the heart and rapid angiographic images.

Echocardiogram (Echo) Heart USS. Local referral procedures vary.

- **2-dimensional** Produces a fan-shaped, cross-sectional, moving, real-time image of the heart. May be transthoracic or transoesophageal. Used to asses valvular abnormalities and prosthetic heart valves; aortic aneurysm/dissection; heart failure; pericardial effusion; masses within the heart; myocardial abnormalities (e.g. aneurysms, hypertrophy); IHD; congenital heart disease

- **M-mode** Plotted on a scrolling screen. Stationary structures appear as straight lines across the screen; moving structures appear as undulating lines. Usually displayed with an ECG trace to enable identification of phases of the cardiac cycle. Used to investigate movement of individual structural elements, e.g. valves, chamber walls
- **Doppler** Enables flow across valves and ASD/VSDs to be quantified

Cardiac enzymes Biochemical blood assay of molecules released when the heart is damaged. Used in diagnosis of MI.

- **Troponins T and I** Preferred markers as more sensitive/specific than CK, AST, or lactate dehydrogenase. Together with CK, earliest to ↑ after MI
- **Creatine kinase (CK)** ↑ in MI, muscle damage, e.g. prolonged running or seizures, after IM injection, and with dermatomyositis (e.g. due to statins). CK-MB assay may help clarify whether a cardiac event has occurred <48h previously
- **AST** 2nd to ↑
- **Lactate dehydrogenase (LDH)** Last to ↑

Cardiac catheterization Refer via 2° care. Involves passing a catheter, usually via the femoral or brachial artery, to the heart. Used to:

- Measure pressures within the heart and great vessels
- Assess oxygen saturation via blood samples
- Perform coronary angiography—contrast is injected into the coronary arteries to assess their anatomy and/or patency
- Perform intravascular ultrasound
- Perform other procedures, e.g. angioplasty, valvuloplasty, cardiac biopsy

Complications Arrhythmia (0.56%); MI (0.07%); stroke (0.07%); death (0.14%); haemorrhage at the site of insertion (0.56%); thromboembolism; trauma to heart and vessels; infection.

Exercise ECG ECG testing while the patient undergoes graded exercise on a treadmill/exercise bicycle. Mortality ~1 in 10,000. Used for:

- Diagnosis of IHD—although cardiac MRI is now the preferred test. For patients with IHD, 75% have a +ve exercise test; false +ve rate of ~5%
- Assessment of exercise tolerance
- Response to treatment
- As a prognostic indicator
- Assessment of exercise-related arrhythmias

Contraindications Recent MI (<7d), unstable angina, electrolyte disturbance, aortic stenosis, severe heart failure, known left main coronary artery stenosis, LBBB (may not be possible to interpret the trace).

Radionucleotide imaging 2° care tests. Involves IV administration of a γ-emitting radionucleotide and gamma camera monitoring.

- **Radionucleotide angiography** Uses technetium^{99m}-labelled RBCs to calculate left ventricular ejection fraction/assess ventricular action
- **Myocardial perfusion scintigraphy** Uses thallium^{201} injected IV during exercise testing to demonstrate areas of poorly perfused myocardium

Patient information

British Heart Foundation ☎ 0300 330 3311 🖱 www.bhf.org.uk

Brief guide to common ECG changes

❗ For detailed analysis of ECGs refer to a specialist text (e.g. Hampton J et al. *The ECG Made Easy* (9th edition—2019) Churchill Livingstone, ISBN: 9780702046414).

Table 9.4 Common ECG abnormalities and their causes

ECG abnormality		Possible causes
Tachycardia	Rate >100bpm	Physiological, AF, atrial flutter, SVT, VT
Bradycardia	Rate <60bpm	Physiological, drugs (e.g. β-blockers, digoxin), heart block (see ➔ p. 242), sick sinus syndrome
Irregular	Assess whether any pattern or not	AF (no pattern), sick sinus syndrome (no pattern), ventricular ectopics (normally no pattern), heart block (pattern)
P–R interval	Short P–R interval	Nodal rhythm, WPW syndrome (➔ p. 239)
	Prolonged >0.2sec	Heart block—➔ p. 242; sick sinus syndrome, drugs (e.g. β-blockers, digoxin)
*Left bundle branch block (LBBB)**	QRS >0.12sec wide. Last peak is below the isoelectric line in V1	IHD, ↑ BP, cardiomyopathy, aortic valve disease, SVT. Artificial pacemakers may produce a similar QRS complex
Right bundle branch block (RBBB)	QRS >0.12sec wide. Last peak is above the isoelectric line in V1	May be normal; congenital heart disease (e.g. ASD), valvular heart disease, IHD, pulmonary hypertension, during SVT
Incomplete bundle branch block	QRS <0.12sec with abnormal shaped QRS complex	As for RBBB or LBBB
Q–T interval abnormalities	Prolonged Q–T interval	↓ K^+, drugs (e.g. TCAs, phenothiazines, amiodarone), SAH or CVA, hypothermia, genetic
	Shortened Q–T interval	↑ Ca^{2+}, digoxin
Abnormal P waves	↑ P-wave amplitude (>2.5mm)	Right atrial overload—tricuspid stenosis, pulmonary hypertension, pulmonary stenosis
	Biphasic P wave in V1 ± broad (>0.12sec) often notched P wave in ≤1 limb lead	Left atrial abnormality—mitral stenosis, aortic stenosis, conduction abnormalities
Right ventricular hypertrophy (RVH)	Strain pattern—ST depression and T-wave inversion in leads V1–3. Dominant R in V1 with narrow QRS	Pulmonary stenosis, mitral stenosis pulmonary hypertension, ASD (± RBBB). Similar changes seen with inferior MI (T-wave upright); WPW syndrome

* No comment can be made about ST segment or T wave if LBBB.

(*Continued*)

Table 9.4 (*Contd.*)

ECG abnormality		Possible causes
Left ventricular hypertrophy (LVH)	Strain pattern—ST ↓ and T wave ↓ in leads V4–6. Large voltages of QRS complex—sum of S in V1 and R in V5 or V6 alone >35mm	↑ BP, aortic stenosis, coarctation of the aorta, HOCM
Right axis deviation	➔ p. 210	RVH/strain (e.g. following PE), cor pulmonale, pulmonary stenosis. Alone with normal QRS = left posterior hemiblock
Left axis deviation	➔ p. 210	LVH /strain (e.g. ↑ BP, aortic stenosis, HOCM), VSD, ASD. If occurs alone with normal QRS = left anterior hemiblock
Poor R-wave progression	Absence of the normal ↑ in size of the R wave in the precordial leads from V1 to V6	Old anterior MI; lead misplacement (common in obese women); LBBB or left anterior fascicular block; LVH; WPW syndrome; dextrocardia; tension pneumothorax with mediastinal shift; congenital heart disease
Abnormal Q waves	>25% of succeeding R wave and/or >0.04sec wide	Normal; left pneumothorax; dextrocardia; MI; myocarditis; hyperkalaemia; cardiomyopathy; amyloid; sarcoid; scleroderma; LVH; RVH; LBBB; WPW syndrome
ST elevation	ST segment raised >1mm above baseline	MI, Prinzmetal angina, pericarditis, ventricular aneurysm
ST depression	ST segment lowered >0.5mm below baseline	Angina, ventricular strain, drugs (digoxin, verapamil), hyperkalaemia, myocarditis, cardiomyopathy, fibrosis, Lyme disease
T-wave inversion	Abnormal if inverted in leads I, II or V4–6	MI (inverts <24h after MI); ventricular strain (see above); PE (III); digoxin (V5–6)
U-waves	↑ amplitude >1mm	Drugs (e.g. quinidine, procainamide, disopyramide) or ↓ K^+
Inversion in precordial leads		Subtle sign of ischaemia

❗ Always compare with previous ECGs if available.

Prevention of cardiovascular disease

1 in 3 deaths result from heart disease; in England alone, there are ~32,000 stroke-related deaths each year in England.

Risk factors for cardiovascular disease Table 9.5

Primary prevention Aims to stop cardiovascular disease developing.

Population strategy Influences factors that ↑ CHD risk in an entire population, e.g. anti-smoking campaigns. GPs can do this by displaying health education posters/literature (e.g. in waiting room, practice leaflet).

Health checks for the over 40s There is a national screening programme for people aged 40–74y in the UK to identify CVD, DM, and cancer earlier through health checks every 5y. The check provides:

- Lifestyle advice (➔ p. 223)
- Information about cancer screening programmes available in the UK
- CVD risk assessment—including demographic information, FH, smoking status, alcohol consumption, BMI, BP, and lipid profile
- DM risk stratification—➔ p. 315

High-risk strategy Most CVD occurs in individuals of medium risk as that is the largest group. However, individuals at high risk have the most to gain from risk reduction. This strategy aims to identify individuals at high risk through opportunistic screening and health checks and ↓ their risk through lifestyle modification ± medication. High-risk groups:

- All people aged >85y
- All patients with a familial dyslipidaemia—➔ p. 225
- People with eGFR <60 mL/min/1.73m^2 and/or albuminuria
- People with type 1 DM who are >40y or have had DM for >10y or have established nephropathy or other CVD risk factors
- People with 10y CVD risk of ≥10% using the QRisk3 calculator (🖱 https://qrisk.org/three/)

❗ Current risk estimation tools give 10y CVD risk; the Joint British Societies tool provides an estimate of lifetime risk (🖱 www.jbs3risk.com). This is particularly useful when discussing CVD risk with younger patients as their 10y risk is often very low regardless of risk factors.

Factors used in the QRisk3 cardiovascular risk calculation

- Age
- Gender
- Ethnicity
- Socioeconomic status
- BMI
- Cholesterol levels
- Smoking
- Diabetes
- FH of CVD
- AF
- BP
- BP medication
- CKD
- RA
- SLE
- Severe mental illness
- Antipsychotic medication
- Steroid medication

❗ If the risk score is near the threshold for intervention, consider other factors that may predispose to CVD and cause risk underestimation, e.g.:

- Has the person recently stopped smoking?
- Is the patient already taking lipid-lowering therapy?
- Does the patient have raised triglycerides?
- Are other conditions present that ↑ risk (e.g. psoriasis, HIV, other systemic inflammatory disorder, taking immunosuppressant medication)?

Secondary prevention Aims to stop progression of symptomatic cardiovascular disease. 46% people who die from MI are already known to have CHD. There is strong evidence that targeting patients with CVD for risk-factor modification is effective in ↓ risk of recurrent CVD.

Table 9.5 Risk factors for cardiovascular disease

Non-modifiable	Modifiable (proven benefit)	Modifiable (unproven benefit)
Age—↑ with age Sex—♂ > ♀ in those < 65y Ethnic origin—in the UK people who originate from the Indian subcontinent have ↑ risk, Afro-Caribbeans have ↓ risk Socioeconomic position* Personal history of CVD Family history of CVD—<55y ♂; <65y ♀ Low birth weight (IUGR)	Smoking*—➔ p. 156 Hyperlipidaemia—➔ p. 222 Hypertension*—➔ p. 218 DM*—➔ p. 326 Diet*—➔ p. 148 Obesity (particularly waist-hip ratio)*—➔ p. 152 Physical inactivity*—➔ p. 154 Alcohol consumption*—➔ p. 158 Left ventricular dysfunction/heart failure (2° prevention)—➔ p. 234 Coronary prone behaviour—competitiveness, aggression and feeling under time pressure (2° prevention)—behaviour modification is associated with ↓ risk	Haemostatic factors—↑ plasma fibrinogen Apolipoproteins—↑ lipoprotein(a)* Homocysteine—↑ blood homocysteine Vitamin levels—↓ blood folate, vitamins B_{12} and B_6 Depression

* These 9 factors account for 90% of risk for acute MI.

The GP's role GPs have a role in:

- Identification of patients who would benefit from primary prevention
- Ensuring patients at high risk of atherosclerotic disease have ongoing follow-up through disease registers, routine recall, and follow-up
- Promoting lifestyle modification in at-risk patients (➔ p. 223)
- Ensuring current best care guidelines are followed and treatment regimens are updated as policies change; this is a major challenge for primary care as guidelines change frequently and often conflict
- Checking the process through audit

Further information

NICE (2014, updated 2016) Cardiovascular risk assessment and the modification of blood lipids for the primary and secondary prevention of cardiovascular disease. ☞ www.nice.org.uk/guidance/cg181
SIGN (2017) Risk estimation and the prevention of cardiovascular disease. ☞ https://www.sign.ac.uk/sign-149-risk-estimation-and-the-prevention-of-cardiovascular-disease.html

Patient information

British Heart Foundation ☎ 0300 330 3311 ☞ www.bhf.org.uk

Blood pressure measurement

Taking blood pressure

- Use a validated sphygmomanometer—a list is available on the British Hypertension Society website (🖱 www.bhsoc.org)
- Regularly maintain and calibrate your sphygmomanometer
- Use a cuff of correct bladder size: most adults—12 × 26cm; large adults (arm circumference >33cm)—12 × 40cm; thin adults and children with arm circumference ≤26cm—10 × 18cm
- Seat the patient comfortably with arm outstretched and supported at the level of the heart
- Measure BP to the nearest 2mmHg; measure diastolic BP when heart sounds completely disappear (K_5)—only use the pressure at which they suddenly muffle (K_4) when K_5 cannot be determined
- Measure BP in both arms—if the difference in systolic BP is >10mmHg between the 2 arms, repeat the measurement. A consistent difference of >10mmHg is an independent risk factor for CVD and all-cause mortality; treat identified CVD risk factors. Use the higher of the 2 readings when considering further management of BP

❗ Automated devices may not measure BP accurately if there is pulse irregularity (e.g. AF). Check the radial or brachial pulse before checking BP and measure BP manually using direct auscultation over the brachial artery if pulse is irregular.

'White coat' hypertension Prevalence 10%. Some patients' BP increases in response to having their BP checked

If BP is ≥140/90mmHg in the consultation

- Take a 2nd measurement—if that measurement is substantially different from the 1st measurement, take a 3rd measurement; record the lower of these measurements as the BP
- If BP on repeat testing is ≥140/90mmHg and not known to be hypertensive, offer ambulatory BP monitoring (ABPM) or home BP monitoring to confirm the diagnosis; consider ABPM or home BP monitoring (HBPM) for people on antihypertensive medication who are known to have 'white coat' hypertension
- If hypertension is not confirmed on ABPM/HBPM, measure BP every 5y or more frequently if close to 140/90mmHg

⚠ If the person has severe hypertension (systolic BP ≥180mmHg or diastolic BP ≥110mmHg), consider starting antihypertensive treatment immediately without waiting for the result of ABPM/HBPM.

Refer for same-day specialist assessment if suspected:

- Accelerated hypertension (BP>180/110mmHg ± papilloedema ± retinal haemorrhage) or
- Phaeochromocytoma (labile/postural hypotension, headache, palpitations, pallor, and excessive sweating)

Ambulatory BP monitoring (ABPM) A BP measuring device is worn on the body for 24h. It takes BP every 20min in the day time and less frequently (usually hourly) at night. ↑ BP readings on ABPM are more strongly correlated to end-organ damage than one-off measurements. ABPM is used for:

- The diagnosis of hypertension (➔ p. 218)

- Diagnosis or exclusion of 'white coat' hypertension
- Monitoring of response to treatment
- Assessing medication-related postural hypotension

Interpretation ABPM is considered abnormal if
- Average day-time ABPM is ≥135/85mmHg
- Average night-time ABPM is ≥120/70mmHg

❗ NICE bases the diagnosis of hypertension on average ***day-time*** ABPM only (minimum 14 readings).

Night-time ABPM BP falls at night in normotensive individuals. Some hypertensive patients are described as 'non-dippers'. This means that their BP does not fall at night and is associated with ↑ risk of target end-organ damage. Conversely, some hypertensive patients have excessive dips in their BP at night (>10%); this is associated with ↑ CVD events.

Home BP monitoring (HBPM) Useful:
- If ABPM facilities are not available within a reasonable time frame for diagnosis of hypertension
- For patients who cannot tolerate ABPM
- For monitoring of antihypertensive treatment

Instructions Patients should use a validated and calibrated BP machine.
- Measure BP on at least 4—and preferably 7—consecutive days
- Measure BP 2×/d (morning and evening) when seated; take 2 readings >1min apart
- Discard the readings taken on day 1 and take an average value of all the remaining measurements

Interpretation HBPM is abnormal if average is ≥135/85mmHg.

Hypertension ➔ p. 218 **Cardiogenic shock** ➔ p. 1065

Postural hypotension BP drops on moving from supine or sitting position to standing position. Presents with falls, postural dizziness, and/or light-headedness. Measure BP supine or seated and then ask the patient to stand up. Re-measure BP after 1min standing. Standing usually causes a slight ↓ in the systolic BP (<20mmHg) and a slight ↑ in the diastolic BP (<10mmHg). In postural hypotension there is usually a marked ↓ in both systolic (>20mmHg) and diastolic BP.
- Review medication Stop any non-essential medication contributing to symptoms, e.g. night sedation, unnecessary diuretics
- Optimize treatment of intercurrent heart disease, Parkinson's disease, or DM
- Advise patients to take care when standing—especially if getting up from their beds or out of a hot bath/shower, and after meals

Measure subsequent BP with the patient standing. Consider specialist referral if symptoms persist despite GP management.

Further information

Clark CE, et al. (2012) The difference in blood pressure readings between arms and survival: primary care cohort study. *BMJ* **344**:e1327.

NICE (2011, updated 2016) Hypertension in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg127

Hypertension

Hypertension is a major risk factor for CVD. It is normally symptomless until it causes organ damage. Prevalence ↑ with age; 25% of all adults and >50% of people aged >60y have hypertension. Management aims to detect and treat ↑ BP before damage occurs.

Causes

- Unknown ('essential')—95%; alcohol (10%) or obesity may be contributory factors
- Endocrine disease—Cushing's (both syndrome and 2° to steroids); Conn's syndrome; phaeochromocytoma; acromegaly; hyperparathyroidism; DM
- Renal disease
- Pregnancy
- Coarctation of the aorta—➔ p. 252

Presentation

- Usually asymptomatic and found during routine BP screening or incidentally. Occasionally headache or visual disturbance
- May be symptoms of end-organ damage—LVH, TIAs, previous CVA/MI, angina, renal impairment, PVD

Measurement of blood pressure ➔ p. 216

Diagnosis of hypertension BP is a normally distributed continuous variable; each 2mmHg ↑ in systolic BP is associated with a 7% ↑ risk of mortality from IHD and a 10% ↑ risk of mortality from stroke. There is no figure above which hypertension can be diagnosed definitively. Criteria currently in use[N]:

- **Stage 1 hypertension** Clinic BP ≥140/90mmHg and subsequent daytime average ABPM/HBPM ≥135/85mmHg
- **Stage 2 hypertension** Clinic BP ≥160/100mmHg and subsequent daytime average ABPM/HBPM ≥150/95mmHg
- **Severe hypertension** Clinic systolic BP ≥180mmHg or clinic diastolic BP ≥110mmHg

⚠ If the person has severe hypertension (systolic BP ≥180mmHg or diastolic BP ≥110mmHg), consider starting antihypertensive treatment immediately without waiting for the result of ABPM/HBPM.

Refer for same day specialist assessment if suspected:

- Accelerated hypertension (BP >180/110mmHg ± papilloedema ± retinal haemorrhage) or
- Phaeochromocytoma (labile/postural hypotension, headache, palpitations, pallor, and excessive sweating)

Isolated systolic hypertension Systolic BP ≥160mmHg—offer the same treatment as people with ↑ systolic and diastolic BP.

Further assessment Aims to identify target organ damage:

- **Examination** Check heart size, heart sounds, and for heart failure; examine the fundi, looking for silver wiring, AV nipping, flame haemorrhages and cotton wool spots
- **Blood tests** Creatinine, electrolytes, eGFR, glucose/HbA1c, lipid profile, consider GGT if excess alcohol is a possibility

- **Urine** Dipstick for RBCs and protein; laboratory sample for albumin: creatinine ratio
- **Cardiovascular risk estimation** ➔ p. 214
- **ECG ± Echo** (if LVH is suspected)

❗ If ↑ BP is not diagnosed but there is evidence of target organ damage (e.g. LVH, proteinuria), look for alternative causes.

Education Patients will not take tablets regularly, be motivated to change lifestyle, or turn up for regular checks if they do not understand why treating their ↑ BP is important. Conversely, some patients who were fit and well prior to their diagnosis will assume a sick role unless it is explained that they are well and treatment is designed to stop illness developing. Back-up verbal information with written information that patients can take home; reinforce information at follow-up and offer opportunities for discussion. Do not forget to warn patients about possible side effects of any medications.

Lifestyle advice Offer lifestyle advice to all patients with a diagnosis of hypertension and those with FH of ↑ BP. Reinforce advice with written information:

- Offer smoking cessation advice and help (➔ p. 156)
- ↓ weight to optimum for height (➔ p. 152)
- Encourage regular exercise—dynamic is best, e.g. walking, swimming, cycling (➔ p. 154)
- ↓ alcohol to <14U/wk for ♂ and ♀ (➔ p. 158)
- ↓ dietary salt intake (aim for <6g salt/d)
- ↑ dietary fruit and vegetable intake—aim for >5 portions/d
- ↓ excess coffee consumption and other caffeine-rich products
- Encourage relaxation and stress management
- *Do not* offer Ca^{2+}, Mg^{2+}, or K^+ supplements as a method to ↓ BP

Statin (➔ p. 224). Prescribe:

- If hypertension complicated by CVD irrespective of baseline cholesterol or LDL levels *or*
- For primary prevention in patients >40y with hypertension and 10y CVD risk ≥10%

Initiating antihypertensive drug treatment

Stage 1 hypertension Offer drug treatment if <80y of age and ≥1 of:

- Target organ damage
- Established CVD
- Renal disease
- DM
- 10y CVD risk ≥10%

Stage 2 hypertension Offer drug treatment to all patients.

❗ If aged <40y, stage 1 hypertension, and no evidence of target organ damage, CVD, renal disease, or DM, consider specialist referral to exclude 2°causes of hypertension and for detailed estimation of CVD risk.

General rules for antihypertensive drug treatment

- **If a drug is not tolerated** Stop; move to the next line of therapy
- **If a drug is tolerated but the BP target is not met** Add in the next line of therapy
- **Where possible** Recommend treatment with drugs taken only once a day and prescribe non-proprietary drugs which minimize cost

Choice of antihypertensive drug(s) Figure 9.1 ❗ Side effects of drug treatment for hypertension are common. 40–50% started on an antihypertensive drug discontinue regardless of which class of drug is used. 80% of side effects are seen in 1st year of treatment.

Treatment targets

Non-diabetic patients without CKD

- Clinic BP <140/90mmHg (<150/90mmHg if aged ≥80y)
- ABPM/HBPM (e.g. for patients with white coat hypertension) average daytime BP <135/85mmHg (<145/85mmHg if aged ≥80y)

Patients with diabetes (➜ p. 326)

- <140/80mmHg—uncomplicated type 2 DM
- <135/85mmHg—uncomplicated type 1 DM
- <130/80mmHg—if any renal, foot, eye or cardiovascular complications of type 1 or type 2 DM

CKD Aim for BP <140/90mmHg; <130/80mmHg if CKD + DM or CKD + ACR of ≥70mg/mmol (➜ p. 415).

History of stroke/TIA Aim for systolic BP <130mmHg unless carotid stenosis when target is 140–150mmHg.

Follow-up Regular review of patients with ↑ BP is essential.

Review interval Depends on stability of BP:

- After starting treatment Review after 1mo
- If BP is controlled Review after a further 3mo, then every 3–6 mo
- If BP is not controlled Bring the patient back to repeat the BP reading and/or ask for ABPM/HBPM. *Do not* alter medication on the strength of a single BP reading. If ↑ BP is sustained, alter medication. Review in the same way monthly until BP is controlled

Format of the annual review

- Check BP and look for signs of end-organ failure—including annual urine test for proteinuria
- Discuss symptoms and medication
- Assess and treat other modifiable risk factors for CHD/CVA
- Reinforce lifestyle advice

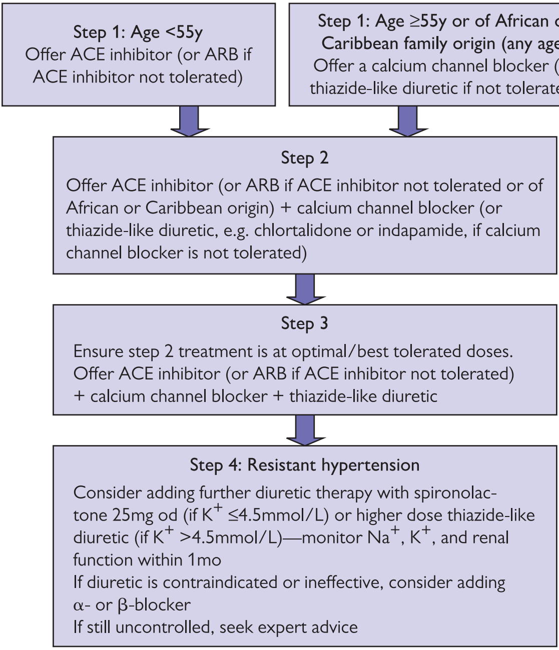
Referral to cardiology/general medicine

E = Emergency admission; *U* = Urgent; *S* = Soon; *R* = Routine.

- Accelerated hypertension—*E*
- Renal impairment—*U/S/R*
- Suspected secondary hypertension—*U/S/R*
- Patients <40y or BP difficult to treat (step 4)—*R*
- Pregnancy—to obstetrician—urgency depends on stage of pregnancy and clinical features—➜ p. 804

Reducing or stopping treatment ↓ BP too far (<120/80mmHg) may ↑ morbidity (e.g. as a result of postural hypotension and falls)—particularly in the elderly:

- *Do not* stop medication if high CVD risk or end-organ damage
- If diastolic BP <80mmHg and systolic BP <140mmHg consistently, consider ↓ or stopping medication; 1–2y after withdrawal of medication 50% are normotensive and 40% stay off drug therapy permanently
- Continue BP follow-up lifelong even if off medication



Step 1: Age <55y
Offer ACE inhibitor (or ARB if ACE inhibitor not tolerated)

Step 1: Age ≥55y or of African or Caribbean family origin (any age)
Offer a calcium channel blocker (or thiazide-like diuretic if not tolerated)

Step 2
Offer ACE inhibitor (or ARB if ACE inhibitor not tolerated or of African or Caribbean origin) + calcium channel blocker (or thiazide-like diuretic, e.g. chlortalidone or indapamide, if calcium channel blocker is not tolerated)

Step 3
Ensure step 2 treatment is at optimal/best tolerated doses.
Offer ACE inhibitor (or ARB if ACE inhibitor not tolerated) + calcium channel blocker + thiazide-like diuretic

Step 4: Resistant hypertension
Consider adding further diuretic therapy with spironolactone 25mg od (if K^+ ≤4.5mmol/L) or higher dose thiazide-like diuretic (if K^+ >4.5mmol/L)—monitor Na^+, K^+, and renal function within 1mo
If diuretic is contraindicated or ineffective, consider adding α- or β-blocker
If still uncontrolled, seek expert advice

Figure 9.1 Choice of antihypertensive drug

β-blockers β-blockers are not recommended as initial therapy for hypertension. *Exceptions*:

- Younger women of child-bearing potential
- Patients with hypertension and evidence of ↑ sympathetic drive
- Patients intolerant of or with contraindications to ACE inhibitors/ARBs

If initial therapy is with a β-blocker and a second drug is required, add a non-rate-limiting calcium channel blocker to ↓ risk of DM.

Management of hypertension in pregnancy ➔ p. 804

Further information

NICE (2011, updated 2016) Hypertension in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg127

NICE (2014, updated 2016) Cardiovascular risk assessment and the modification of blood lipids for the primary and secondary prevention of cardiovascular disease. 🖱 www.nice.org.uk/guidance/cg181

Patient information

British Heart Foundation ☎ 0300 330 3311 🖱 www.bhf.org.uk

Hyperlipidaemia

Average cholesterol level in a population is a predictor of CVD risk and dependent on diet but, on an individual level, it is a much poorer predictor—only 42% who develop CVD have ↑ cholesterol. However, lowering LDL and raising HDL is thought to reduce progression of coronary atherosclerosis, whatever the age of the patient, and is a valuable tool for both primary and secondary prevention of CVD.

Cholesterol testing Cholesterol is a fatty substance manufactured by the body (mainly liver) which plays a vital role in functioning of cell membranes. Cholesterol levels are checked with a non-fasting blood sample[N]. A lipid profile includes:

- **HDL (high-density lipoprotein) cholesterol**—low levels are associated with ↑ risk CVD
- **Non-HDL cholesterol**—low-density lipoprotein (LDL) + very low-density lipoprotein (VLDL). High levels are associated with ↑ CVD risk
- **Triglycerides (TGs)**—independent risk factor for CVD
- **Ratio of total cholesterol:HDL**—used to predict risk. No threshold—the higher the ratio the greater the risk. High risk if ≤6

! Consider familial dyslipidaemia (➔ p. 225) in any patient if total cholesterol >7.5mmol/L and FH of premature CVD. Refer for specialist assessment if total cholesterol >9.0mmol/L or non-HDL cholesterol >7.5mmol/L, regardless of FH[N].

Management of raised triglycerides[N]

- If TGs >20mmol/L—refer for urgent specialist review if not a result of excess alcohol or poor glycaemic control
- If TGs 10–20mmol/L—repeat the triglyceride measurement with a fasting test (after an interval of ≥5d, but in <2wk). Review for potential secondary causes of hyperlipidaemia (Box 9.1) and seek specialist advice if TGs remain ≥10mmol/L
- If TGs 4.5–9.9mmol/L—CVD risk may be underestimated by risk assessment tools. Optimize management of other CVD risk factors. Seek specialist advice ↑ TGs + non-HDL cholesterol >7.5mmol/L

Lipid screening In the UK, the NHS provides health checks every 5y for those aged 40–74y (➔ p. 214). These include a lipid profile. Potential problems with this strategy:

- Those most needing a health check are least likely to attend
- There may be less inclination to eat a healthy diet if cholesterol levels are known to be normal
- Those with ↑ cholesterol levels may assume a sick role

Blood cholesterol concentration is also not steady over time. 1 in 4 ↑ cholesterol levels are normal on repeat testing. If lipid levels are high on screening, repeat with attention to diet; only treat with a statin if lipids remain high despite low-cholesterol diet and 10y CVD risk is ≥10%.

Opportunistic screening Offer if other risk factors for CVD or signs of ↑ cholesterol (e.g. corneal arcus <50y, xanthelasma, xanthomata).

Calculating cardiovascular disease (CVD) risk Always consider cholesterol in the context of other risk factors for CVD—➔ p. 215.

Secondary hyperlipidaemia High lipid levels as a consequence of another condition—Box 9.1.

Box 9.1 Secondary causes of hyperlipidaemia

Drugs:
- Steroids
- β-blockers
- Thiazides
- COC pill
- Isotretinoin
- Antipsychotics
- Tamoxifen
- Antiretrovirals

Obesity
DM (➔ p. 314)[a]
Excess alcohol
Smoking (lowers HDL)
Pregnancy
Hypothyroidism[b]
Renal failure
Nephrotic syndrome
RA/SLE

HIV
Cholestasis
Cushing's syndrome
Porphyria
Myeloma
Lipodystrophies
Glycogen storage disease

[a] Treatment of hyperglycaemia in DM ↓ secondary hyperlipidaemia.

[b] Patients with hypothyroidism should receive adequate thyroid replacement before assessing need for lipid-lowering treatment. Correction of hypothyroidism may resolve the lipid abnormality and untreated hypothyroidism ↑ risk of myositis with statins.

Non-drug therapy Offer to all patients with ↑ cholesterol, ↑ CVD risk, and those with DM or personal/family history of CHD/CVD. Reinforce advice with written information.

- ↓ intake of fats to <30% of total energy intake (saturated fats <7%) and ↓ cholesterol intake to <300 mg/d. Replace saturated fats with monounsaturated/polyunsaturated fats. If cholesterol level >5mmol/L, low-cholesterol diets result in ↓ in cholesterol of 8.5% at 3mo
- Eat ≥5 portions of fruit or vegetables/d, 4–5 portions unsalted nuts, seeds, or legumes/wk, and ≥2 portions of fish/wk (including 1 of oily fish—maximum 2 portions of oily fish/wk if pregnant); choose wholegrains; reduce sugar
- ↓ alcohol intake to ≤14U/wk; <3–4U/d (♂) or <2–3U/d (♀) maximum
- Weight ↓—in patients with BMI ≥30kg/m^2, weight ↓ of 10kg → 7% ↓ in non-HDL and 13% ↑ in HDL cholesterol
- ↑ physical activity—enhances cholesterol-lowering effects of diet and weight ↓. Advise 150min moderate (or 75min vigorous) exercise/wk + muscle strengthening exercises 2×/wk
- Stop smoking (➔ p. 156)

❗ Plant sterol/stanol esters inhibit cholesterol absorption and can ↓ serum cholesterol with average diet by 10%. ↓ effect if already on a low-fat diet. Avoid if on lipid-lowering medications, DM (type 1 or 2) or CKD.

Who should be treated with a statin? Consider statin therapy for:

Primary prevention If:
- >85y (if appropriate)
- 10y CVD risk ≥10%
- eGFR <60mL/min/$1.73m^2$ and/or albuminuria
- Type 1 DM + >40y or DM for >10y or established nephropathy or other CVD risk factors

Start treatment after optimizing lifestyle intervention, and treatment of other modifiable risk factors/secondary causes of dyslipidaemia.

Secondary prevention If history of CVD—start treatment immediately irrespective of initial cholesterol levels

Factors to consider before starting a statin

- Discuss risks vs benefits. Consider other co-morbidities, polypharmacy, general frailty, and life expectancy before deciding to prescribe
- Contraindicated in pregnancy, breastfeeding, and for those with active liver disease (transaminases ≥3× upper limit of normal)
- Drug interactions—statins ↑ effect of warfarin; there is ↑ risk myositis when statins are taken with other lipid-lowering drugs, macrolide antibiotics (e.g. erythromycin), antifungals (e.g. ketoconazole), amiodarone, calcium channel blockers, HIV protease inhibitors (e.g. nelfinavir), danazol, or ciclosporin
- Diet restrictions—avoid grapefruit if taking atorvastatin, simvastatin, or lovastatin, and cranberry juice (fluvastatin only)

Baseline assessments before starting a statin

- Smoking status
- Alcohol consumption
- BP
- BMI
- Lipid profile
- Liver function (transaminases)
- HbA1c
- Renal function
- eGFR
- TSH

Only measure creatine kinase (CK) level if history of unexplained persistent, generalized muscle pain.

- If CK <5× upper limit of normal, start statin at ↓ dose
- If ≥5× upper limit of normal, recheck after 7d—if still ≥5× upper limit of normal, do not start a statin

Starting dose Statins are most effective taken in the evening. Use a high-intensity statin (↓ LDL by >40%):

- **Primary prevention** Start atorvastatin 20mg after optimizing lifestyle intervention, and treatment of other modifiable risk factors
- **Secondary prevention** Start atorvastatin 80mg immediately

> ❗ Rosuvastatin 10–40mg is an alternative. Simvastatin 80mg (also a high-intensity statin) should not be commenced due to ↑ risk of myopathy.

Target lipid levels Aim to ↓ non-HDL by >40% from baseline.

Follow-up

- Recheck lipid profile in 3mo. If target is not met: promote lifestyle measures, check concordance, ↑ dose (if tolerated/not on maximum)
- Check liver function 3mo and 1y after initiating statin/changing dose; do not recheck again unless clinically indicated
- Do not routinely check CK levels—only if muscle pain

Benefits of statins ↓ CVD risk overall by ~25%. In general, the higher the risk of CVD at baseline, the greater the benefit of statins. Therefore, for all patients with established CVD benefit of statin treatment (in terms of ↓ in CVD risk) greatly outweighs risk.

Primary prevention As overall risk is lower in primary prevention, benefit of statin treatment is not so clear cut. Decision aids may help patients to decide whether or not they wish to take a statin. Options:

- NICE—provides a paper-based decision aid in the 'tools and resources' section of its lipid modification guidance
- QRisk3 and JBS3 risk calculator tools can both be used to graphically demonstrate risk reduction likely for individual patients from modification of lifestyle factors and/or taking statins—➔ p. 214

Adverse effects of statins Overall NNH for high-intensity statin therapy is ~63. If unable to tolerate a statin, consider ↓ dose, or prescribing an alternative statin or ezetimibe. Specific side effects:

- Myositis Most important adverse effect of statins (11/100,000 person years). Risk is ↑ if elderly or alcohol misuse. Usually arises <6mo after starting treatment—consider other causes if later. May be genetic predisposition. Ask patients to report unexplained muscle pain/weakness—check CK—if >5× upper limit of normal withdraw therapy
- Peripheral neuropathy Stop statins and seek specialist advice if unexplained peripheral neuropathy develops (12/100,000 person years)
- Abnormal liver function Discontinue if serum transaminase ↑ and remains at >3× upper limit of normal
- Diabetes ↑ risk type 2 DM. Consider screening with HbA1c

Alternative lipid-lowering treatments Do not offer fibrates, nicotinic acid, bile acid sequestrants, or omega-3 fatty acid for prevention of CVD. Ezetimibe may be used if statin is contraindicated/not tolerated, or combined with low-dose statin to achieve target cholesterol levels.

Familial dyslipidaemia Screen 1st-degree blood relatives aged >18y every 5y with fasting lipids if FH of familial hyperlipidaemia and/or FH of premature CVD (♂ <55y, ♀ <65y). Exclude secondary causes of hyperlipidaemia (Box 9.1, ➔ p. 223). Refer if[N]:

- Total cholesterol >7.5mmol/L and FH of premature CVD
- Total cholesterol >9mmol/L, non-HDL cholesterol >7.5mmol/L; or TGs >10mmol/L on repeat fasting sample (urgently if >20mmol/L)

Common types of familial dyslipidaemia

- Polygenic hypercholesterolaemia Most common. Presents with FH of premature CHD + ↑ total cholesterol >6.5mmol/L
- Familial combined hyperlipidaemia Polygenic. Affects 0.5–1% of the population and ~15% having MI <60y. Associated with obesity, insulin resistance/DM, ↑ BP, xanthelasma, corneal arcus and premature IHD. ↑ total cholesterol (6.5–10mmol/L); ↑ TGs (2.3–12mmol/L)
- Familial hypercholesterolaemia Autosomal dominant. Heterozygous form affects 1 in 500 individuals. Associated with tendon xanthomata and FH premature IHD. ↑ LDL (>4.9mmol/L); ↑ total cholesterol (>7.5mmol/L); normal TGs
- Familial hypertriglyceridaemia Autosomal dominant. Affects ~1% of the population and ~5% having MI <60y. Associated with DM, obesity, gout, eruptive xanthomas, and pancreatitis. Normal (or slightly ↑) total cholesterol; ↑ TGs (2.3–>10mmol/L)

Further information

NICE (2014, updated 2016) Cardiovascular risk assessment and the modification of blood lipids for the primary and secondary prevention of cardiovascular disease. 🖱 www.nice.org.uk/guidance/cg181

NICE (2008, updated 2017) Familial hypercholesterolaemia. 🖱 www.nice.org.uk/guidance/cg71

SIGN (2017) Risk estimation and the prevention of cardiovascular disease. 🖱 https://www.sign.ac.uk/sign-149-risk-estimation-and-the-prevention-of-cardiovascular-disease.html

Patient information

British Heart Foundation ☎ 0300 330 3311 🖱 www.bhf.org.uk

Angina

Affects 8% ♂ and 3% ♀ in the UK. Incidence ↑ with age. Coronary artery disease is the most common cause. Rarer causes include HOCM, valve disease, hypoperfusion during arrhythmia, arteritis, anaemia or thyrotoxicosis. Mortality (usually sudden death, acute coronary syndrome, or LVF) is ~0.5–4%/y—doubled if coexistent left ventricular dysfunction.

Chest pain of recent onset ➲ p. 1064

Presentation of stable angina Diagnosis is usually made on history:

- **Pain** Episodic central-crushing or band-like chest pain that may radiate → jaw/neck and/or 1 or both arms. Pain in the arms/neck may be the only symptom. Ask about frequency, severity, duration, and timing
- **Precipitating/relieving factors** Precipitated by exertion, cold, emotion, and/or heavy meals. Pain stops with rest or GTN spray
- **Associated symptoms** Pain may be associated with palpitations, sweating, nausea, and/or breathlessness during attacks
- **Presence of risk factors** Smoking history; family history; history of other vascular disease, e.g. CVA/TIA, peripheral vascular disease

Examination There are usually no physical signs although anaemia may exacerbate symptoms. Check BMI and BP. Look for murmurs (especially ejection systolic murmur of aortic stenosis) and evidence of peripheral vascular disease and carotid bruits (especially in patients with DM).

First-line investigations

- **Blood** FBC, fasting lipid profile, fasting blood glucose. Consider checking ESR (to exclude arteritis) and TFTs if suspicion of thyrotoxicosis
- **12-lead resting ECG** Provides information on rhythm, presence of heart block, previous MI, myocardial hypertrophy and/or ischaemia

❗ A normal ECG does not exclude coronary artery disease, but an abnormal ECG identifies those at higher risk of cardiac events in the next year—consider referral for further investigation.

Differential diagnosis Chest pain—➲ p. 1065

Referral of patients with suspected stable angina For patients with new-onset intermittent chest pains, refer to a rapid access chest pain clinic for prompt specialist assessment to:

- Confirm/refute angina
- Perform appropriate investigations, e.g. cardiac MRI (➲ p. 210)
- Provide information on treatment options available including the merits of revascularization for the individual

❗ Patients with pre-existing cardiac disease (e.g. previous MI, valve disease, cardiomyopathy) are often excluded from rapid access chest pain clinic referral. Refer to cardiology direct.

Management of patients with stable angina Provide information about angina and its treatment.

- **Driving** *Group 1 licence*: no need to inform DVLA—must not drive when symptoms occur at rest, with emotion or at the wheel. *Group 2 licence*: notify the DVLA—must not drive when symptoms occur; licence is revoked if symptoms continue; restored if no angina for ≥6wk + DVLA functional test requirements are met

- **Occupation** May be unable to undertake heavy work—give advice/support. Special rules apply to some occupations, e.g. merchant seamen, airline pilots. Advise to consult occupational health

Non-drug treatment Aimed at secondary prevention of CHD:

- **Smoking cessation** ➔ p. 156
- **Hypertension** Check BP and treat if >140/90mmHg—➔ p. 218
- **Diet** Advise healthy diet (oily fish, low cholesterol, ↑ fruit and vegetables, ↓ salt) and, if obese, aim to ↓ weight until BMI <25kg/m^2
- **↓ alcohol** To ≤14U/wk; <3–4U/d (♂) or <2–3U/d (♀) maximum
- **Exercise** ↑ aerobic exercise within the limits set by the disease state; consider referral for cardiac rehabilitation
- **Diabetes** Treat any underlying DM—➔ p. 320
- **Cardiac rehabilitation** May be helpful for patients with angina and/or after surgery

Drug treatment ➔ p. 228

Referral to cardiology *E*= Admit; *U* = Urgent; *S* = Soon; *R* = Routine.

- Unstable angina/rapidly progressive symptoms—*E*
- Aortic stenosis with angina—*U*
- Angina following MI—*U/S*
- Abnormal ECG at diagnosis—*U/S*
- Angina not controlled by medication with 2 drugs—*U/S/R*
- If diagnosis is in doubt—*S/R*
- Strong family history—*R*
- Other factors, e.g. occupation affected—*R*

⚠ **Unstable angina** Pain on minimal or no exertion, pain at rest (may occur at night) or angina which is rapidly worsening in intensity, frequency, or duration. *Incidence*: 6/10,000/y; 15% suffer MI in <1mo.

Management Urgent referral to cardiology. Admit if attacks are severe, occur at rest or last >10min even with GTN spray.

Surgical intervention Consider referral for coronary revascularization with bypass surgery (CABG) or percutaneous intervention (PCI) if symptoms are not controlled with antianginal drug intervention (2 drugs). Both procedures ↓ symptoms, but CABG confers a survival advantage if DM, age >65y, left anterior descending (LAD) artery disease, or complex 3-vessel disease.

Prinzmetal (variant) angina Angina at rest resulting from coronary artery spasm. ECG shows ST elevation. Refer to cardiology to exclude MI and atherosclerotic angina. GTN alleviates immediate episodes. Calcium channel blockers are used to prevent angina.

Cardiac syndrome X Ongoing angina symptoms despite normal coronary angiography. Treat with β-blockers and/or calcium channel blockers (if effective) but not secondary prevention agents.

Further information

Cardiac rehabilitation 🖱 www.cardiacrehabilitation.org.uk

NICE (2011, updated 2016) Stable angina: management. 🖱 www.nice.org.uk/guidance/cg126

Patient information

British Heart Foundation ☎ 0300 330 3311 🖱 www.bhf.org.uk

Drug treatment of angina

Symptom control

'As required' medication Glyceryl trinitrate (GTN) spray is used for 'as required' symptom relief for angina. Advise 1–2 puffs as needed in response to pain and before engaging in activities that bring on pain. Warn about side effects—flushing, headaches, and light-headedness (sit down or find something to hold onto if this occurs).

> ⚠ If used for chest pain: advise patients to repeat the dose of GTN after 5min if the pain has not gone. If the pain has not gone 5min after the second dose, advise to call for an emergency ambulance.

Regular treatment Drugs for regular symptomatic treatment—Table 9.6. Within any drug class, use the cheapest preparation that the patient can tolerate, will comply with, and which controls symptoms. Assess response every 2–4wk after initiating/changing drug therapy:

- **First-line agent** β-blocker or calcium channel blocker—choice depends on co-morbidities, contraindications, and patient preference
- **If treatment is ineffective/not tolerated** Switch to whichever first-line agent has not been tried and/or combine a β-blocker and dihydropyridine calcium channel blocker

Alternative regular treatments Include long-acting nitrates, ivabradine, nicorandil, and ranolazine. Consider:

- Monotherapy if both first-line agents (β-blockers and calcium channel blockers) are contraindicated or not tolerated
- In combination with a first-line agent if symptoms are not controlled with one first-line agent alone and the other first-line agent is contraindicated or not tolerated
- As a third antianginal agent if symptoms are not controlled with 2 antianginal drugs and the person is either not suitable for CABG/PCI or is awaiting CABG/PCI

Secondary prevention

Aspirin ↓ mortality by 34%. Unless contraindicated, give 75mg od to *all* patients with angina. Consider clopidogrel 75mg od if aspirin intolerant.

Statin A reduction in total cholesterol and LDL by 25–35% results in a reduction of CHD mortality by 25–35%. Trial data suggest *all* patients with proven CHD benefit from statin treatment irrespective of initial cholesterol concentration—➔ p. 223.

ACE inhibitors Significantly ↓ cardiovascular deaths (RR 0.83) and all-cause mortality (RR 0.87)—even in the absence of left ventricular dysfunction.

Table 9.6 Drug treatment of angina

Drug	Treatment notes
β-blockers ❗ May accumulate in patients with renal failure—↓ dose	Effective for symptom control and to prevent vascular events. Check fully β-blocked by monitoring heart rate—resting heart rate ≤65bpm; post-exercise (e.g. walking up 2 flights of stairs) heart rate ≤90bpm. Further ↑ in dose once adequately β-blocked are usually unhelpful Warn patients not to stop suddenly or run out. If the patient needs to stop the drug, tail off over 4wk In patients with asthma/COPD in whom β-blockade is essential, use cardio-selective β-blockers (e.g. atenolol, bisoprolol, metoprolol, nebivolol) with care In patients with left ventricular failure, start at very low dose and titrate dose over weeks/months
Dihydropyridine calcium channel blockers	e.g. amlodipine, felodipine All equally effective in symptom control. No evidence of cardioprotective effect *Contraindications*: vary. Do not use if aortic stenosis, <1mo post-MI or uncontrolled heart failure except with specialist advice
Rate-limiting calcium channel blockers	e.g. diltiazem, verapamil *Contraindications*: avoid if heart block or heart failure ⚠ Do not combine with β-blockers
Long-acting nitrates	e.g. isosorbide mononitrate (ISMO) Oral and patch preparations (dosages ≤ 10mg/24h) are available. Start with a low dose and ↑ as tolerated. Side effects are common *Side effects*: headache, postural hypotension and dizziness—wear off with use. Reflex tachycardia may ↓ coronary blood flow and worsen angina *Tolerance*: many patients rapidly develop tolerance with ↓ therapeutic effect. To avoid this allow a nitrate-free period of 4–8h/d overnight by removing patches at night or giving the 2nd dose of ISMO at 4 p.m. *Contraindications*: HOCM, aortic stenosis, constrictive pericarditis, mitral stenosis, severe anaemia, closed-angle glaucoma
Potassium channel activator	e.g. nicorandil Headache is common—usually transitory *Contraindications*: left ventricular failure; hypotension
Ivabradine	Lowers the heart rate by its action on the sinus node *Contraindications*: avoid if heart rate <60bpm, heart block, or heart failure
Ranolazine	Affects sodium-dependent calcium channels *Contraindications*: renal/liver failure; use with caution if CCF or weight <60kg

Further information

NICE (2011, updated 2016) Stable angina: management. 🖱 www.nice.org.uk/guidance/cg126

Patient information

British Heart Foundation ☎ 0300 330 3311 🖱 www.bhf.org.uk

After myocardial infarction

Acute coronary syndrome ➔ p. 1066

Modification of risk factors after MI Secondary prevention:

- Cholesterol All with proven CHD benefit from ↓ in total cholesterol and LDL irrespective of initial cholesterol level. A reduction of 25–35% using statin therapy results in 25–35% ↓ in CHD mortality. Serum cholesterol levels ↓ after MI and remain ↓ for several weeks
- β-blockers Unless contraindicated, start on an oral β-blocker (e.g. atenolol) soon after MI and titrate to target or maximum tolerated dose. Continue for 12mo, or indefinitely if LVF. Prevents ~12 deaths /1000 treated/y. If contraindicated, consider a rate-limiting calcium channel blocker (e.g. diltiazem or verapamil) instead

ACE inhibitors ↓ myocardial work and deaths <1mo post MI by 5/1000 treated. Titrate to target or maximum tolerated dose in 4–6wk. Survival advantage is sustained >1y even if treatment is not continued. Effects are greater for patients with heart failure at presentation. *Long-term ACE inhibitors*: trials show ↓ mortality for all patients. If ACE inhibitors are not tolerated, use an ARB.

Antiplatelet medication

Aspirin Starting aspirin 75mg od <24h after MI prevents 80 vascular events over the next 2y/1000 patients treated. Unless contraindicated, continue lifelong.

Clopidogrel 75mg od—prescribe in addition to aspirin for:

- 12mo if non-ST elevation MI (NSTEMI)/unstable angina
- 1mo following ST elevation MI (STEMI) with no coronary stenting
- 3mo following STEMI with bare-metal stenting
- 12mo following STEMI with drug-eluting stenting

Alternatives to clopidogrel In certain circumstances, newer antiplatelet agents (e.g. prasugrel or ticagrelor) may be used in combination with aspirin after ACS and PCI.

Anticoagulation Occasionally required if AF, left ventricular aneurysm, or if clopidogrel/aspirin are not tolerated.

Drug treatment of angina ➔ p. 228

Heart failure/left ventricular dysfunction Consider treatment with an aldosterone antagonist (e.g. spironolactone 25mg od, ↑ in <1mo to 50mg od), starting 3–14d after MI if symptoms/signs of heart failure and left ventricular systolic dysfunction (ejection fraction <0.4).

Cardiac rehabilitation ↓ risk of death by 20–25%. Provided by specialist multidisciplinary teams. *Components include*: psychological support, information about CHD, structured exercise programme, and modification of other risk factors.

Advice to provide after discharge

Physical activity Advise gradual ↑ in activity. Ensure goals given match those given by local cardiac rehabilitation. Guide:

- Up to 2wk—stroll in the garden or street

- 2–6wk—walk ½ mile/d aiming to ↑ to 2 miles/d by 6wk
- From 6wk—↑ speed of walking—aim 2 miles in <30min

Sexual activity Resume as comfortable—usually after ≥4wk. A leaflet is available from the British Heart Foundation.

Return to work Guide:

- Sedentary workers: 4–6wk after uncomplicated MI
- Light manual workers: 6–8wk after uncomplicated MI
- Heavy manual workers: 3mo after uncomplicated MI

Psychological effects ~50% are depressed 1wk after MI; 25% after 1y. Educate about CHD. Check for depression (➔ p. 173), counsel, and treat as needed.

Driving Group 1 licence No need to notify the DVLA. Stop driving. Restart after 1wk if successful PCI and no other revascularization planned in <4wk. Otherwise driving can restart 4wk after the event.

Driving Group 2 licence Inform the DVLA. Licence is revoked; reinstated after ≥6wk if LV ejection fraction is ≥40% + DVLA functional test requirements are met.

Flying Most airlines will not carry passengers for 2wk after MI and then only if able to climb 1 flight of stairs without difficulty.

Ongoing GP follow-up

Monitoring health Continue regular reviews at least annually lifelong. Check for symptoms and signs of cardiac dysfunction (breathlessness, palpitations, angina); depression; carer stress.

Monitoring drug therapy Ongoing prescription of drugs, monitoring of compliance and side effects, changing medication if clinical circumstances or best practice alter.

Secondary prevention

- Smoking cessation ➔ p. 156. ↓ risk of death by 50% over 15y
- Hypertension Check BP and treat if >140/90mmHg—➔ p. 218
- Diet Mediterranean-style diet (low cholesterol, ↑ fruit/vegetables, ↓ salt) and, if obese, aim to ↓ weight until BMI <25kg/m^2. 💣 Do not advise oily fish to ↓ risk of further MI (but no evidence of harm)
- ↓ alcohol to ≤14U/wk; <3–4U/d (♂) or <2–3U/d (♀) maximum
- Exercise ↑ aerobic exercise within the limits set by the disease state
- Diabetes Treat any underlying DM—➔ p. 320
- Reinforce information given during cardiac rehabilitation

Dressler syndrome (post-MI syndrome) Develops 2–10wk after MI or heart surgery as a result of autoantibodies to heart muscle. Presents with recurrent fever and chest pain ± pleural and/or pericardial effusion. *Management*: Refer urgently for cardiology advice. Treatment is with steroids and NSAIDs.

Further information

Cardiac rehabilitation 🖱 www.cardiacrehabilitation.org.uk

NICE (2013) Myocardial infarction: cardiac rehabilitation and prevention of further cardiovascular disease. 🖱 www.nice.org.uk/guidance/cg172

Patient information

British Heart Foundation ☎ 0300 330 3311 🖱 www.bhf.org.uk

Chronic heart failure

Chronic heart failure occurs when output of the heart is inadequate to meet the body's needs. It is the end stage of all diseases of the heart. 900,000 people in the UK have heart failure; prevalence ↑ with age.

Acute heart failure ➔ p. 1068

Causes of chronic heart failure

High-output The heart is working at normal or ↑ rate but the needs of the body are ↑ beyond that which the heart can supply, e.g. hyperthyroidism, anaemia, Paget's disease, AV malformation.

Low-output ↓ heart function. Causes:

- **↑ pre-load** e.g. mitral regurgitation, fluid overload
- **Pump failure**:
  - Cardiac muscle disease—IHD (46%), cardiomyopathy
  - ↓ expansion of the heart and restricted filling—restrictive cardiomyopathy, constrictive pericarditis, tamponade
  - Inadequate heart rate—β-blockers, heart block, post MI
  - Arrhythmia—AF is the most common
  - ↓ power—negatively ionotropic drugs, e.g. verapamil, diltiazem
- **Chronic excessive afterload** ↑ BP, aortic stenosis

Presentation Clinical diagnosis is difficult. Take a detailed history and do a clinical examination to exclude other disorders.

Algorithm for diagnosing heart failure Figure 9.2

Differential diagnosis

- Obesity
- Respiratory disease
- Venous insufficiency in lower limbs
- Drug-induced ankle swelling (e.g. calcium channel blockers) or fluid retention (e.g. NSAIDs)
- Intrinsic renal or hepatic disease
- Pulmonary embolic disease
- Hypoalbuminaemia
- Depression and/or anxiety
- Bilateral renal artery stenosis
- Severe anaemia
- Thyroid disease

Causes of falsely ↑ natriuretic peptide

- Structural or functional cardiac disease of any cause, including MI
- Baseline is ↑ in ♀ and in those >70y
- Lung disease, including COPD and PE
- Renal impairment
- DM
- Liver failure
- Sepsis

Causes of falsely ↓ natriuretic peptide

- Obesity
- Diuretics
- ACE inhibitors/ARBs
- β-blockers
- Aldosterone antagonists

Classification

- **Left ventricular systolic dysfunction** ↓ left ventricular ejection fraction (LVEF) on echocardiography
- **Heart failure with preserved ejection fraction (HFPEF)** Also termed diastolic dysfunction—signs/symptoms of heart failure with normal LVEF on echocardiogram

Other tests to consider To exclude aggravating factors/other causes of symptoms—urinalysis, blood (FBC, U&E, creatinine, eGFR, TFTs, FBG/HbA1c), ECG, CXR, and PEFR/spirometry.

Grading of severity The New York Heart Association (NYHA) classification is widely used:

- I No limitation: ordinary physical exercise does not cause undue fatigue, dyspnoea, or palpitations
- II Slight limitation of physical activity: comfortable at rest but ordinary activity results in fatigue, palpitations, or dyspnoea
- III Marked limitation of physical activity: comfortable at rest but less than ordinary activity results in symptoms
- IV Unable to carry out any physical activity without discomfort: symptoms of heart failure are present even at rest with ↑ discomfort with any physical activity

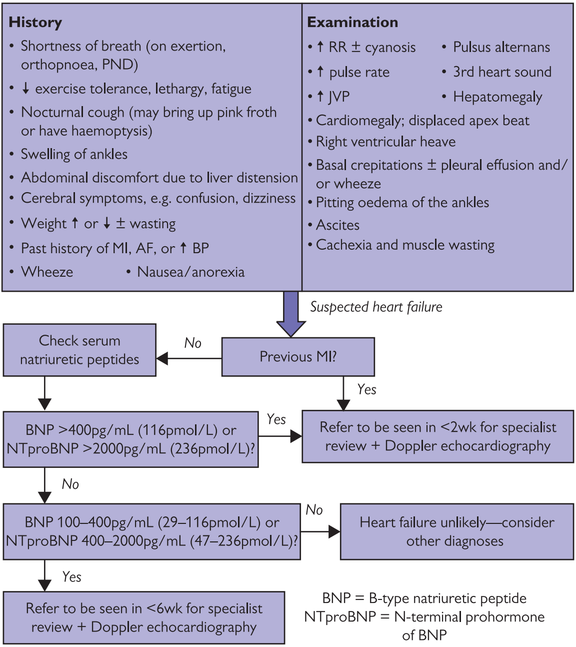

History
• Shortness of breath (on exertion, orthopnoea, PND)
• ↓ exercise tolerance, lethargy, fatigue
• Nocturnal cough (may bring up pink froth or have haemoptysis)
• Swelling of ankles
• Abdominal discomfort due to liver distension
• Cerebral symptoms, e.g. confusion, dizziness
• Weight ↑ or ↓ ± wasting
• Past history of MI, AF, or ↑ BP
• Wheeze
• Nausea/anorexia
Examination
• ↑ RR ± cyanosis
• ↑ pulse rate
• ↑ JVP
• Pulsus alternans
• 3rd heart sound
• Hepatomegaly
• Cardiomegaly; displaced apex beat
• Right ventricular heave
• Basal crepitations ± pleural effusion and/or wheeze
• Pitting oedema of the ankles
• Ascites
• Cachexia and muscle wasting
Suspected heart failure
Previous MI?
No
Check serum natriuretic peptides
Yes
Refer to be seen in <2wk for specialist review + Doppler echocardiography
BNP >400pg/mL (116pmol/L) or NTproBNP >2000pg/mL (236pmol/L)?
Yes
No
BNP 100–400pg/mL (29–116pmol/L) or NTproBNP 400–2000pg/mL (47–236pmol/L)?
No
Heart failure unlikely—consider other diagnoses
Yes
Refer to be seen in <6wk for specialist review + Doppler echocardiography
BNP = B-type natriuretic peptide
NTproBNP = N-terminal prohormone of BNP

Figure 9.2 Algorithm for diagnosing heart failure

Further information

NICE (2018) Chronic heart failure in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng106

Patient information

British Heart Foundation ☎ 0300 330 3311 🖱 www.bhf.org.uk

Management of chronic heart failure

❗ Always look for the underlying cause and treat wherever possible. Review the basis for historical diagnosis and arrange Echo to confirm if diagnosis is in doubt.

Driving and heart failure

- **Group 1 licence** Stop driving and inform the DVLA if severe symptoms (NHYA IV)
- **Group 2 licence** Inform the DVLA. Can continue driving if mild symptoms (NYHA I or II) and LV ejection fraction is >40%

Regular review Every 6mo or more often as needed. Check:

- **Clinical state** Functional capacity, fluid status, cardiac rhythm, cognitive and nutritional status
- **Screen for depression** Affects >40%
- **Manage co-morbidities**
- **Medication** Ensure drug record is up to date, review compliance and side effects, change if clinical circumstances/best practice alter
- **Blood** U&E, creatinine, and eGFR

Non-drug measures

- **Educate** about the disease, current/expected symptoms, and need for treatment. Discuss prognosis. Support with written information
- **Discuss ways to make life easier** e.g. benefits, mobility aids, blue disability parking badge. Consider referral to social services for assessment for services such as home care
- **Diet** Adequate calories, ↓ salt, ↓ weight if obese, restrict alcohol
- **Lifestyle measures** Smoking cessation (➔ p. 156); regular exercise
- **Restrict fluid intake** If severe heart failure
- **Vaccination** Pneumococcal and annual influenza vaccination

Diuretics Relieve congestive symptoms/fluid retention in all types of heart failure. Choose a loop diuretic, e.g. furosemide 20–40mg or bumetanide 1–2mg od. Add a thiazide if continued problems with oedema or hypertension. Titrate dose ↑ or ↓ according to need. Monitor for ↓ K^+ and co-treat with amiloride or K^+ supplements as needed.

First-line medication for left ventricular systolic dysfunction Start all patients on an ACE inhibitor *and* a β-blocker. Use clinical judgement to decide which drug to start first.

ACE inhibitors Improve symptoms, ↑ exercise capacity, ↓ progression of disease, ↓ hospital admissions, and ↑ survival in symptomatic and asymptomatic patients. Start at low dose (e.g. ramipril 1.25mg od) and titrate upwards. Check U&E and Cr before starting, at first follow-up and after each ↑ in dose. Use ARB if not tolerated.

❗ If neither an ACE inhibitor nor ARB is tolerated first-line, a combination of hydralazine with a nitrate is an alternative—seek specialist advice.

β-blockers Start a β-blocker licensed for heart failure (e.g. bisoprolol 1.25mg in the morning) in all those with left ventricular dysfunction regardless of whether symptoms persist. Use in a 'start low, go slow' manner with assessment of pulse, BP, and clinical status after each titration.

Heart failure with preserved ejection fraction (HFPEF) Apart from diuretics, there is no specific treatment for HFPEF. Treat any co-morbidities, e.g. DM, ↑ BP, IHD.

Other drugs to consider

- **Anticoagulation** If heart failure + AF, or history of thromboembolism, left ventricular aneurysm, or intra-thoracic thrombus—➔ p. 648
- **Aspirin** 75–150mg od if heart failure + atherosclerotic arterial disease (including CHD)
- **Statins** Only if other indications—➔ p. 224
- **Amlodipine** Treatment for angina and ↑ BP. ❗ Avoid verapamil, diltiazem, or short-acting dihydropyridine agents

Referral to cardiology (or other suitable specialist) Consider if (*E* = Emergency admission; *U* = Urgent; *S* = Soon; *R* = Routine):

- Heart failure unable to be managed at home—*E*
- Severe heart failure—*U/S*
- Heart failure not controlled by first-line medication—*U/S/R*
- Angina, AF, or other symptomatic arrhythmia—*U/S/R*
- Heart failure due to valve disease or diastolic dysfunction—*R*
- Co-morbidity that may impact heart failure, e.g. COPD, renal failure, anaemia, thyroid disease, PVD, urinary frequency, gout—*R*
- Woman with heart failure planning pregnancy—*R*

Treatment under specialist supervision

Left ventricular systolic dysfunction Second-line agents include:

- Aldosterone antagonists, e.g. spironolactone
- Combination of hydralazine and nitrate
- ARB—used in combination with ACE inhibitor and β-blocker

Digoxin An antiarrhythmic and a positive inotrope. It is used for worsening or severe heart failure due to left ventricular systolic dysfunction despite first- and second-line treatment. Only check levels (8–12h after last dose) if suspected toxicity or non-adherence.

Amiodarone May be used to treat arrhythmias associated with heart failure. It requires specialist initiation and close monitoring with TFTs and LFTs at least every 6mo once established on a maintenance dose.

Medical devices

- **Implantable cardioverter defibrillator** May be fitted in patients with left ventricular dysfunction and previous episodes of ventricular tachycardia or widened QRS complexes on ECG
- **Cardiac resynchronization therapy** Considered if severe symptoms on maximal therapy and in sinus rhythm with prolonged QRS complexes

Prognosis Progressive deterioration to death; ~½ die suddenly—probably due to arrhythmias. *Mortality*: Mild/moderate heart failure—20–30% 1y mortality; severe heart failure—>50% 1y mortality

Palliative care ➔ p. 1011

Further information

NICE (2018) Chronic heart failure in adults: management. 🖱 www.nice.org.uk/guidance/ng106

Patient information

British Heart Foundation ☎ 0300 330 3311 🖱 www.bhf.org.uk

Pulmonary hypertension and cor pulmonale

Pulmonary hypertension Normal pulmonary arterial pressure is less than a fifth of that in the systemic circulation. Pulmonary hypertension is defined as pulmonary artery pressure ≥25mmHg. It is classified according to cause:

- **Group 1** Pulmonary arterial hypertension—may be idiopathic or associated with connective tissue disease (mainly scleroderma)
- **Group 2** Left heart disease—severe heart failure (systolic or diastolic), valve disease (e.g. mitral stenosis), congenital heart disease
- **Group 3** Lung disease—COPD, interstitial lung disease
- **Group 4** Chronic thromboembolic pulmonary hypertension—PE, sickle cell disease
- **Group 5** Unclear and multifactorial mechanisms

Cor pulmonale Right heart failure resulting from chronic pulmonary hypertension.

Diagnosis Non-specific symptoms often lead to delayed diagnosis.

Presentation Fatigue and breathlessness on exertion ± angina, palpitations, and pre-syncope. Later, as ventricular failure develops, symptoms include peripheral oedema, ascites, and syncope.

Examination Check for cyanosis, peripheral oedema, ↑ JVP, 4th heart sound, diastolic murmur from pulmonary regurgitation, hepatomegaly ± ascites, crepitations at lung bases ± pleural effusion.

Investigations

- **CXR** Prominent right heart border + enlargement of proximal pulmonary arteries
- **ECG** If abnormal—right axis deviation, tall peaked P wave in lead II, dominant R wave in V1, T-wave inversion in anterior leads or RBBB

Management Refer to a specialist cardiologist or chest physician.

Specialist investigation

- **Doppler Echo** Used to assess ventricular function and give an estimate of pulmonary arterial pressure
- **Right heart catheterization** and direct measurement of mean pulmonary artery pressure Confirms diagnosis, provides information on prognosis, and determines the treatment plan
- **Assessment of functional capacity** The 6min walk test is used to give information about prognosis; <380m at presentation is associated with poor prognosis

Specialist treatment Depending on the underlying cause, treatment is usually through a tertiary specialist centre and includes:

- Oxygen therapy for symptomatic relief
- Diuretic treatment for heart failure
- Vasodilation with calcium channel blockers (in the 10–15% of patients who are responsive)
- Anticoagulation

- Specific drug treatment (group 1 pulmonary hypertension)
- Surgery

Specific drug treatments For group 1 pulmonary hypertension. Improve symptoms, delay disease progression, and prolong survival. Always initiated in specialist centres:

- Prostanoids e.g. epoprostenol, iloprost, selexipag
- Endothelin receptor antagonists e.g. bosentan, ambrisentan, macitentan—require monthly monitoring with LFTs
- Phosphodiesterase inhibitors e.g. sildenafil, tadalafil
- Other agents e.g. riociguat (causes arterial dilatation by action on an enzyme that mediates the vasomotor effect of nitric oxide in the lungs)

Pulmonary endarterectomy Treatment of choice for patients with cardio-thromboembolic pulmonary hypertension. The majority can be cured with normalization of pulmonary pressure following surgery (>90% 5y survival).

Bilateral heart/lung transplantation For those who have severe symptoms despite optimal treatment. 5y survival following transplantation is 50–60%.

Driving and heart failure ➔ p. 234

Patient information

Pulmonary Hypertension Association (PHA) UK ☎ 01709 761450
🖱 www.phauk.org

Tachycardia

Tachycardia is a heart rate >100bpm. It is often felt as palpitations. Common and may be an incidental finding. History and examination can exclude significant problems in most patients.

Driving and arrhythmia ➔ p. 241

History Ask about:

- Palpitations Duration, frequency and pattern, rhythm (ask the patient to tap it out if not present when seen)
- Precipitating/relieving factors
- Associated symptoms Chest pain, collapse or funny turns, sweating, breathlessness or hyperventilation
- Past history e.g. previous episodes, heart disease, thyroid disease
- Family history e.g. any cardiac/rhythm disorders, sudden death
- Lifestyle Drug history; caffeine/alcohol intake; smoking
- Occupation Arrhythmias may affect driving and/or work

⚠ Red flag symptoms

- Pre-existing cardiovascular disease
- FH of syncope, arrhythmia, or sudden death
- Arrhythmia associated with falls and/or syncope

Examination

- General examination For anaemia, thyrotoxicosis, anxiety, other systemic disease
- Cardiovascular examination Heart size, pulse rate and rhythm, JVP, BP, heart sounds and murmurs, evidence of left ventricular failure

Investigations Resting ECG is all that is needed for many patients. Consider further investigations if ECG is abnormal or other concerning features (e.g. syncope, breathlessness, prolonged episodes):

- Blood: TFTs, FBC, ESR/CRP, U&E, FBG/HbA1c, Ca^{2+}, albumin
- Ambulatory ECG or cardiac memo
- Echo if <50y or murmur/heart failure detected
- Exercise tolerance test if exercise related

⚠ **Ventricular tachycardia (VT)** Broad (>3 small squares) QRS complexes at a rate of >100bpm on ECG. Admit as a 'blue-light' emergency. Meanwhile give O_2 if available ± 100mg IV lidocaine. If no pulse treat as VF cardiac arrest (➔ p. 1066). If recurrent VT, may require surgery, insertion of a pacemaker or implantable cardioverter defibrillator (ICD).

Ventricular ectopic beats Additional broad QRS complexes, without P waves, superimposed on regular sinus rhythm. Common. Usually of no clinical significance. Rarely, a presenting feature of viral myocarditis.

- Frequent ectopics (>100/h) on ECG Urgent cardiology referral
- R on T phenomenon on ECG Rarely ectopics can lead to VF, particularly if they coincide with the T wave of a preceding beat ('R on T phenomenon'). Admit if this occurs >10×/min on ECG
- After MI Associated with ↑ mortality—refer to cardiology
- No sinister features on ECG Explain benign nature. Advise avoidance of caffeine, alcohol, smoking, and fatigue. β-blockers can be helpful if unable to tolerate ectopics despite reassurance

Long QT syndrome (LQTS) ECG shows prolonged QT interval. Heart is structurally normal. Associated with ventricular arrhythmias which may lead to syncope/sudden death, often triggered by excitement/exercise. If inherited, may be autosomal dominant or recessive. Refer any patient with FH of sudden cardiac death for specialist assessment. Antenatal screening is possible.

Brugada syndrome Particularly affects people of SE Asian origin; ♂ >> ♀. Autosomal dominant inheritance but FH in only ~50%. Results in syncope/sudden death (usually <45y) due to ventricular arrhythmias, often at night. ECG shows 'Brugada' sign—coved ST ↑ of ≥2mm in V1/V2, followed by a −ve T wave—but may be transient/absent. Treatment is with an ICD. Arrange same-day cardiology review if suspected.

Paroxysmal supraventricular tachycardia (SVT) Narrow QRS complex tachycardia with a regular rate >100bpm on ECG.

If seen during an attack Get an ECG if possible. Try carotid sinus massage (unless elderly, IHD, digoxin toxicity, carotid bruit, history of TIAs), the Valsalva manoeuvre, and/or ice on the face (especially effective for children). Admit as an emergency if attack continues.

If attack terminates or not seen during attack SVT may be diagnosed on 12-lead resting ECG or 24h ECG/cardiac memo. Refer to cardiology for assessment and initiation of treatment—urgently if chest pain, dizziness, or breathlessness during attacks. Include ECG if available.

Ongoing care Avoid caffeine, alcohol, and smoking. *Treatment*: catheter ablation or drug therapy (sotalol, diltiazem, verapamil, or flecainide).

Wolff–Parkinson–White (WPW) syndrome A congenital accessory conduction pathway is present between atrium and ventricle (bundle of Kent). Predisposes to SVT and AF. ECG shows a short P–R interval followed by slurred upstroke ('delta wave') into the QRS complex. Refer to cardiology. Treatment is with antiarrhythmics (SVT—verapamil; AF—amiodarone or DC shock) ± ablation of the accessory pathway.

Sinus tachycardia Consider infection, pain, MI, shock, exercise, emotion (including anxiety), heart failure, thyrotoxicosis, drugs.

Inappropriate sinus tachycardia Diagnosis of exclusion. Persistent tachycardia of >95bpm averaged over 24h, with night-time rate dipping. Can result in palpitations, breathlessness, fatigue/exercise intolerance, and syncope/pre-syncope. Cause is unknown. Treatment (if needed) is with β-blockers or rate-limiting calcium channel blockers.

Postural tachycardia syndrome (POTS) Change from lying to standing causes an exaggerated ↑ in pulse rate of >30bpm. ♀ >> ♂. May be a FH. Average age of presentation is 20y. Suspect if palpitations, weakness, and/or syncope/pre-syncope (but no ↓ in BP) on standing. Diagnosis is confirmed by tilt-table testing. Treatments include behavioural modification and drugs, e.g. β-blockers. Cause is unknown. May spontaneously remit.

Atrial fibrillation/flutter ➔ p. 240

No tachycardia and no ECG abnormalities Reassure. Explore the possibility of anxiety disorder (➔ p. 970).

Atrial fibrillation (AF)

Common disturbance of cardiac rhythm characterized by rapid irregularly irregular narrow QRS complex tachycardia with absence of P waves. May be episodic (*paroxysmal*) or chronic. Affects 1.5–2% of the UK population. Prevalence ↑ with age: <0.5% aged 40–50y; 5% >65y; 10% >75y.

Causes Usually idiopathic—may occasionally be due to atrial dilatation (e.g. in association with valve disease, inflammation, or fibrosis of the atrium), or ↑ in muscle mass (e.g. LVH 2° to ↑ BP or cardiomyopathy).

Acute AF May be precipitated by acute infection, high alcohol intake, surgery, electrocution, MI, pericarditis, PE, or hyperthyroidism.

Presentation Often asymptomatic—detected if chance finding of irregular pulse or following stroke/TIA. *If symptomatic*: palpitations, breathlessness, dizziness, postural hypotension, and/or chest discomfort.

Examination Aims to confirm AF and exclude underlying causes. Check pulse rate/rhythm (apex rate > radial pulse rate if in AF), BP, heart size, heart sounds/murmurs, JVP, signs of LVF. Check for signs of systemic disease, e.g. anaemia, thyrotoxicosis.

Assess severity European Heart Rhythm Association (EHRA) Score:
I No symptoms
II Mild symptoms, normal activity not affected
III Severe symptoms, normal activity affected
IV Disabling symptoms, normal activity discontinued

Investigations

- ECG—if paroxysmal AF is suspected but not captured on ECG, consider 24h-ECG monitoring, or cardiac memo
- Blood tests—FBC; U&E/eGFR; TFTs (anaemia, hyperthyroidism or electrolyte imbalance trigger AF; renal function affects drug choice)
- CXR—to evaluate the size of the heart and identify lung disease
- Echocardiography—to check left atrial size and left ventricular function

Management If acutely, severely compromised, admit to hospital.

Rate limitation If the heart is beating too fast, it cannot refill before the next beat and becomes an inefficient pump. Controlling ventricular rate in AF allows the heart to work more effectively. Choice of drugs—Figure 9.3. Offer as the first-line strategy to people with AF, except if:

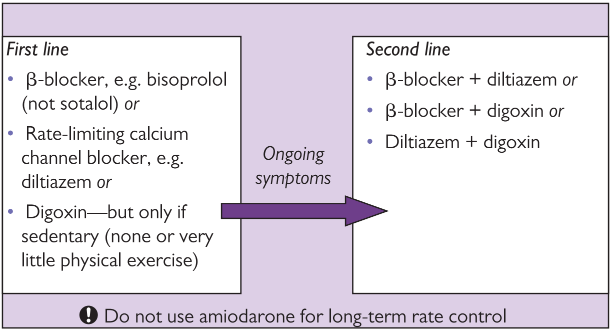
- AF has a reversible cause (e.g. alcohol binge, chest infection)
- Heart failure caused by AF
- New-onset AF
- Atrial flutter suitable for ablation
- Rhythm control would be more suitable

Restoration of sinus rhythm Successful cardioversion is most likely in young patients with a structurally normal heart where duration of AF is <12mo. Consider referral for DC cardioversion if:

- Heart failure associated with AF
- Recent-onset AF with low risk of recurrence, i.e. obvious precipitant (e.g. pyrexia, alcohol, or post-op) and no structural heart disease
- Intolerable symptoms despite adequate rate control
- High risk from anticoagulant therapy

Otherwise offer rate control as the first-line strategy.

❗ Patients are usually started on amiodarone prior to DC cardioversion and for 1y following the procedure to maintain sinus rhythm.



First line

- β-blocker, e.g. bisoprolol (not sotalol) *or*
- Rate-limiting calcium channel blocker, e.g. diltiazem *or*
- Digoxin—but only if sedentary (none or very little physical exercise)

Ongoing symptoms

Second line

- β-blocker + diltiazem *or*
- β-blocker + digoxin *or*
- Diltiazem + digoxin

❗ Do not use amiodarone for long-term rate control

Figure 9.3 Choice of drugs for rate limitation in AF

Prevention of thromboembolic complications ➲ p. 536. AF is associated with an ↑ risk of peripheral thromboembolism including stroke (5× ↑ risk). All patients with valvular AF and moderate/high-risk patients with non-valvular AF should be considered for anticoagulation with warfarin or a direct oral anticoagulant (DOAC, e.g. rivaroxaban, apixaban) to ↓ stroke risk. Do *not* offer aspirin for AF.

Weigh risk of thromboembolism against risk of bleeding (➲ p. 537). The Keele University decision support tool may be useful—🖰 www.anticoagulation-dst.co.uk

'Pill-in-the-pocket' approach to paroxysmal AF Consider self-medication with a β-blocker prn (e.g. atenolol 50–100mg od) if infrequent symptomatic paroxysms *and* no history of LV dysfunction or valvular/ischaemic heart disease; systolic BP >100mmHg and resting heart rate >70bpm; able to understand when and how to take the medication.

Referral to cardiology

E = Emergency admission; *U* = Urgent; *S* = Soon; *R* = Routine

- Acutely unwell with AF—*E*
- For consideration of cardioversion—*E/U*
- Abnormality on echocardiogram requiring referral—*S/R*
- Inadequate rate control using standard drug combinations, still symptomatic despite rate control or frequent attacks of paroxysmal AF—*S/R*
- <30y of age—*S/R*
- Heart failure—*S/R*

Atrial flutter ECG shows regular saw-tooth baseline at rate of 300bpm with a narrow QRS complex tachycardia superimposed at a rate of 100 or 150bpm. Manage as for AF (specialist drug treatment may differ).

Driving and arrhythmia Stop driving if arrhythmia has or is likely to cause incapacity.

- Group 1 licence Restart driving when underlying cause identified and arrhythmia controlled for ≥4wk. Inform the DVLA only if underlying cause not found and arrhythmia not controlled for ≥4wk
- Group 2 licence Inform the DVLA. Licence restored if underlying cause found, arrhythmia controlled ≥3mo + LV ejection fraction >40%

Further information

NICE (2014) Atrial fibrillation: management. 🖰 www.nice.org.uk/guidance/cg180

Bradycardia

Heart rate <60 bpm.

Presentation Often an incidental finding but may present with faints or blackouts, drop attacks, dizziness, breathlessness, or lack of energy.

Examination Slow pulse rate; normal/low BP ± evidence of secondary heart failure. There may also be symptoms/signs of associated disease.

Investigations

- ECG 12-lead resting ECG; ambulatory ECG may help with diagnosis of intermittent bradycardia (e.g. sick sinus syndrome)
- Blood TFTs, FBC, ESR, U&E, LFTs, digoxin levels (if taking digoxin)

Sinus bradycardia Constant bradycardia. P waves present and P–R interval <0.2sec (1 large square). *Causes*:

- Physiological, e.g. athletes
- Vasovagal attack
- Drugs, e.g. β-blockers, digoxin
- Inferior MI
- Sick sinus syndrome
- Hypothyroidism
- Hypothermia
- ↑ ICP
- Jaundice

Management Admit acutely if symptomatic. Refer for cardiology opinion if asymptomatic but HR <40bpm despite treatment of reversible causes.

AV node block (heart block) *Causes* :

- IHD
- Drugs (digoxin, verapamil)
- Myocarditis
- Cardiomyopathy
- Fibrosis
- Lyme disease (rare)

Types of heart block

- 1st-degree block Fixed P–R interval >200msec (1 large square)
- 2nd-degree block:
  - Mobitz type I (Wenckebach)—progressively lengthening P–R interval followed by a dropped beat
  - Mobitz type II—constant P–R interval with regular dropped beats (e.g. 2:1—every second beat is dropped—consider drug toxicity)
- 3rd-degree block (complete heart block)—P–P intervals are constant and R–R intervals are constant but not related to each other

Management Untreated 2nd- and 3rd-degree heart block have a mortality of ~35%. Refer all patients to cardiology even if asymptomatic. If symptomatic (↓ BP <90mmHg systolic, left ventricular failure, heart rate <40bpm) admit as an emergency—give IV atropine and O_2 (if available) while awaiting admission.

Stokes Adams attacks Cardiac arrest due to AV block. Results in sudden loss of consciousness ± some limb twitching due to cerebral anoxia. The patient becomes pale and pulseless but respiration continues. Attacks usually last ~30 sec although occasionally are fatal. On recovery the patient becomes flushed. Refer to cardiology if suspected.

Sick sinus syndrome Due to sinus node dysfunction causing:
- Bradycardia ± asystole
- Sinoatrial block (complete heart block)
- AF or SVT alternating with bradycardia (tachy/brady syndrome)

Common among elderly patients. If symptomatic, heart rate <40bpm or pauses >3sec on ECG, refer to cardiology for pacemaker insertion.

Pacemakers Electrically stimulate the heart to beat. *Indications*:
- Symptomatic bradycardia
- 2nd- or 3rd-degree heart block
- Suppression of resistant tachycardia

Insertion Pacemaker box is attached under the skin of the chest—usually medial to the left axilla—under LA. Wires are fed into the great veins of the chest and thus to the heart under X-ray and/or US guidance.

Types Classified according to:
- Chamber paced—atrium, ventricle, or both ('dual')
- Chamber sensed—atrium, ventricle, or both ('dual')
- Mode of response to sensing—inhibited output, triggered, inhibited, and triggered ('dual')

Thus a VVI pacemaker both paces and senses the ventricle in inhibited mode—i.e. if the ventricle beats spontaneously, the pacemaker will not fire.

ECG changes with a pacemaker If the pacemaker is in operation, a pacing 'spike' (vertical line) is seen on ECG.

❗ In devices pacing on demand, a spike will not be seen if the natural rate is in excess of the rate set on the pacemaker.

Lifespan Pacemakers last 7–15y. Regular checks are made by pacemaker clinics to ensure the pacemaker remains operational. Reprogramming through the skin is possible. Batteries can be changed via a small surgical procedure under local anaesthetic.

Driving and arrhythmia ➔ p. 241

Driving with a pacemaker Inform DVLA and insurance company. Stop driving for 1mo after insertion.

⚠ Pacemakers must be removed after death before cremation can occur. A fee is payable.

Further information

NICE (2010, updated 2014) Transient loss of consciousness (blackouts) in over 16s. 🖱 www.nice.org.uk/guidance/cg109

NICE (2014) Bradycardia: dual chamber pacing. 🖱 www.nice.org.uk/guidance/ta324

Infective endocarditis

⚠ New murmur + fever = endocarditis until proven otherwise.

Infective endocarditis occurs when there is infection of a heart valve (mitral > aortic > tricuspid > pulmonary). The valve may be normal (50%—may be associated with IV drug abuse), rheumatic, degenerative, congenitally abnormal, or prosthetic. Uncommon but consequences may be disastrous and often detected late.

Causes

- **Common organisms** *Strep. viridans* (35–50%); *Staph. aureus* (20%)
- **Non-bacterial causes** SLE, malignancy

Presentation May be acute (acute heart failure) or subacute (course worsening over days/weeks). *Symptoms/signs*:

- **Infective** Fever, weight ↓, night sweats, malaise, lethargy, clubbing, splenomegaly, anaemia, mycotic aneurysms
- **Heart murmurs ± heart failure**
- **Embolic** Stroke, lung abscesses (right heart endocarditis)
- **Vasculitic** Microscopic haematuria, splinter haemorrhages, Osler's nodes (painful lesions on finger pulps), Janeway lesions (palmar macules), Roth's spots (retinal vasculitis), renal failure

High-risk patients Those with:

- Acquired valvular heart disease with stenosis or regurgitation
- Valve replacement
- Structural congenital heart disease, including surgically corrected or palliated structural conditions, but excluding: isolated atrial septal defect, fully repaired ventricular septal defect or fully repaired patent ductus arteriosus, and closure devices that are endothelialized
- Hypertrophic cardiomyopathy
- Previous infective endocarditis

When to suspect infective endocarditis[G] Have a high index of suspicion and admit as an emergency for further investigation if:

- Febrile illness and murmur of new valvular regurgitation
- Febrile illness + pre-existing high-risk cardiac lesion and no clinically obvious site of infection
- Febrile illness associated with any of:
  - Predisposition and recent intervention with associated bacteraemia (e.g. dental work or surgical procedure)
  - Evidence of congestive cardiac failure
  - New conduction disturbance
  - Vascular or immunological phenomena, e.g. embolic event, Roth's spots, splinter haemorrhages, Janeway lesions, or Osler's nodes
  - New stroke
  - Peripheral abscesses (renal, splenic, cerebral, vertebral) of unknown cause
- Protracted history of sweats, weight ↓, anorexia or malaise, and at-risk cardiac lesion
- New unexplained embolic event, e.g. CVA, limb ischaemia

Investigations in primary care if non-acute presentation

- Blood Non-specific signs of infection, e.g. leucocytosis, ↑ ESR/CRP
- ECG 10% develop a conduction defect
- CXR

Management Admit as an emergency if suspected. Avoid starting antibiotics prior to admission as this might cause delay in diagnosis by rendering the blood cultures sterile.

Hospital treatment Once confirmed, treatment is with prolonged IV broad-spectrum antibiotics (≤2wk).

Prognosis 80% have major complications during admission, e.g. heart failure. Valve replacement may be required—especially if endocarditis is on a prosthetic valve. 16–27% die—those with endocarditis affecting a prosthetic valve have poorer prognosis. Other factors predicting poor prognosis:

- Infecting organism—*Staph aureus* 30–40% mortality; streptococci 10% mortality
- ↑ age
- Aortic valve involvement
- Associated heart failure
- CNS complications
- Co-morbidity, e.g. DM

Prevention of infective endocarditis Current guidance advises against routine antibiotic prophylaxis because:

- There is no consistent association between having an interventional procedure and development of infective endocarditis
- Regular toothbrushing presents a greater risk of infective endocarditis than a single dental procedure because of repetitive exposure to bacteraemia with oral flora
- Clinical effectiveness of antibiotic prophylaxis is not proven
- Antibiotic prophylaxis against infective endocarditis may lead to a greater number of deaths through fatal anaphylaxis than a strategy of no antibiotic prophylaxis, and is not cost-effective

Advise instead—about the:

- Importance of maintaining good oral health
- Symptoms that may indicate infective endocarditis and when to seek expert advice
- The risks of undergoing invasive procedures, including non-medical procedures such as body piercing or tattooing

❗ Do not offer chlorhexidine mouthwash as prophylaxis against infective endocarditis to people at risk undergoing dental procedures.

Further information

British Society for Antimicrobial Chemotherapy (2015) Guidelines for the diagnosis and antibiotic treatment of endocarditis in adults. 🖱 www.bsac-arc.com/#details/218

NICE (2008, updated 2016) Prophylaxis against infective endocarditis. 🖱 www.nice.org.uk/guidance/CG64

Rheumatic fever, myocarditis, and pericarditis

Rheumatic fever There has been a dramatic ↓ in incidence of rheumatic fever in industrialized countries since 1950s, but recently numbers of cases have ↑. Rheumatic fever is still an endemic disease in developing countries. *Peak incidence*: age 5–15y.

Cause Rheumatic fever is due to an abnormal immunological response to β-haemolytic streptococcal infection (e.g. 2–4wk after sore throat). Its importance lies in the permanent damage caused to heart valves in some of those affected and subsequent risk of endocarditis.

Diagnosis Can be made if Revised Jones criteria are met (Table 9.7).

Management If suspected refer for specialist care. Specialist management includes evaluation of heart lesions with Echo, bed rest, penicillin, and symptom control (e.g. analgesia, sedatives for chorea). Anti-inflammatory agents such as corticosteroids and aspirin may be used to try to ↓ complications of carditis but their use is controversial.

Prognosis

- 60% develop chronic rheumatic heart disease (70% mitral valve; 40% aortic; 10% tricuspid; 2% pulmonary). Likelihood correlates with severity of initial disease
- Recurrence may occur after further streptococcal infection or be precipitated by pregnancy or combined hormonal contraception

Table 9.7 Revised Jones criteria for diagnosis of rheumatic fever

Requirements for diagnosis of rheumatic fever

Evidence of previous streptococcal infection (scarlet fever, +ve throat swab and/or ↑ ASO titre >200u/mL)
and
2 major criteria
or
1 major + 2 minor criteria

Major criteria	*Minor criteria*
Carditis (45–70%)—arrhythmia, new murmur, pericardial rub, heart failure, conduction defects	Prolonged P–R interval on ECG (but not if carditis is one of the major criteria)
Migratory polyarthritis ('flitting'—75%) red, tender joints	Arthralgia (but not if arthritis is one of the major criteria)
Sydenham's chorea (St Vitus' dance—10%)	Fever
Subcutaneous nodules (2–20%)	↑ ESR or ↑ CRP
Erythema marginatum (2–10%)	History of rheumatic heart disease or rheumatic fever

Source: data from Gewitz MH *et al*. Revision of the Jones Criteria for the Diagnosis of Acute Rheumatic Fever in the Era of Doppler Echocardiography: A Scientific Statement from the American Heart Association. *Circulation*, 131(20), 1806–181, Copyright © 2015 American Heart Association, Inc.

Secondary prevention Penicillin 250mg bd po or sulfadiazine 1g od (500mg od for patients <30kg) for ≤5y to prevent recurrence. Duration of prophylaxis is dependent on whether there was carditis in the initial attack (no carditis—continued for 5y; if cardiac involvement—continued until age 25y or longer).

Acute myocarditis Inflammation of the myocardium. May present in a similar way to MI or with palpitations. *Causes*: viral infection, e.g. Coxsackie virus; diphtheria; rheumatic fever; drugs.

Management Admit for specialist cardiologist care. Treatment is supportive. Some recover spontaneously—others progress to intractable heart failure requiring transplantation.

Pericarditis Sharp, constant sternal pain relieved by sitting forwards. May radiate to left shoulder ± arm or into the abdomen. Worse lying on the left side and on inspiration, swallowing and coughing. A pericardial rub may be present at the left sternal edge on auscultation. *Causes*:

- Infection, e.g. Coxsackie virus, TB
- Malignancy
- Uraemia
- MI (Dressler syndrome ➔ p. 231)
- Trauma
- Radiotherapy
- Connective tissue disease
- Hypothyroidism

Investigations ECG—concave (saddle-shaped) ST elevation in all leads.

Management Refer to cardiology; treat the cause (if possible); symptomatic treatment with NSAID for pain; steroids in resistant cases.

Complications

- **Pericardial effusion** Fluid in the pericardial sac. *Presentation*: heart failure, cardiac tamponade (inability of the heart to dilate in diastole resulting in tachycardia, ↓ BP, ↑ JVP). CXR—large, globular heart. Echo is diagnostic. *Management*: admit for urgent cardiology assessment
- **Constrictive pericarditis** Pericardium becomes fibrosed and non-expansile. Most common cause is TB. *Presentation*: right heart failure, hepatosplenomegaly, ascites, ↓ BP, ↑ JVP. *Management*: refer to cardiologist for confirmation of diagnosis. Treatment involves surgical release of the pericardium

Cardiomyopathy and heart transplant

Cardiomyopathy is primary disease of the heart muscle. Although some cardiomyopathies are ‘unclassified’ most fall into the following 4 groups:

Dilated (congestive) cardiomyopathy Prevalence ≈35/100,000. ♂ > ♀. Dilation of left ± right ventricle and ↓ contractility. Usually presents with heart failure; may also present with arrhythmia, syncope, peripheral embolism, or abnormalities on ECG/Echo. ECG: non-specific S-T abnormalities; CXR: cardiac enlargement and pulmonary venous hypertension. Echo is diagnostic. *Causes*:

- Idiopathic (50%)
- Familial (20%)
- Cardiovascular—IHD, ↑ BP, congenital heart disease, rheumatic heart disease
- Alcohol
- Infection (Coxsackie virus)
- Endocrine disease—myxoedema, thyrotoxicosis, acromegaly
- Cardiotoxic drugs
- Pregnancy
- Connective tissue disease (SLE, PAN, systemic sclerosis)
- Sarcoidosis
- Amyloidosis
- Haemochromatosis
- Malignancy
- Muscular dystrophy

Management Advise patients to stop drinking alcohol as alcohol may make cardiomyopathy worse. Specialist management is needed in all cases and involves:

- Treatment of heart failure and arrhythmias (may require cardiac resynchronization and/or implantable cardiac defibrillator device)
- Most patients require long-term anticoagulation
- Surgery—cardiomyoplasty or heart transplantation

Mortality 40% in 2y (sudden death, cardiogenic shock).

Hypertrophic cardiomyopathy Autosomal dominant inheritance, although 50% are sporadic. In its most common form causes asymmetrical septal hypertrophy ± aortic outflow obstruction (hypertrophic obstructive cardiomyopathy or HOCM).

Presentation Most cases are diagnosed in childhood (<14y) through Echo screening of asymptomatic patients with a FH. *Symptoms/signs*:

- Palpitations—associated with arrhythmias—5% have AF
- Breathlessness on exertion
- Chest pain—may be angina or atypical pain
- Murmur—due to outflow obstruction and/or mitral valve dysfunction
- Faints/collapses

Investigations

- ECG—LVH and ischaemic changes, e.g. T-wave inversion
- CXR—normal until disease is in its late stages
- Echo—diagnostic. Refer if suspicious symptoms or family history

Management and prognosis Ongoing specialist care is essential to provide symptomatic treatment, e.g. β-blockers for chest pain, amiodarone for arrhythmia (digoxin is contraindicated). Implantable cardiac defibrillators improve prognosis for those at high risk of sudden death. Surgical options include septal ablation and myotomy/myectomy to debulk the septum and relieve obstruction.

Mortality Sudden death is unrelated to severity of symptoms.

Restrictive cardiomyopathy Rare. Stiff ventricle limits filling. Presents with heart failure. Echo is diagnostic. *Causes*: amyloid, sarcoidosis, haemochromatosis. *Management*: Specialist management is required. Treatment is symptomatic.

Arrhythmogenic right ventricular cardiomyopathy Rare genetic condition of progressive infiltration of the right ventricular myocardium with fibro-fatty tissue. Often asymptomatic until presents with cardiac arrest. Refer any patients with a suspicious family history.

Family history of sudden death Refer 1st- degree relatives of victims of sudden cardiac death who died aged <45y to cardiology. Antenatal screening for familial cardiomyopathy and LQTS syndrome is possible if familial mutation is known. If FH HOCM and no genetic test:

- Children under <10y—screen with ECG and Echo every 3–5y
- Children aged 10–16y—screen every 6–12mo if there is a family history of HOCM—disease is likely to become apparent at this age
- Young people aged 16–20y—screen annually
- >20y—screen every 5y if FH late-onset hypertrophic cardiomyopathy

❗ Screening intervals are not established for other cardiomyopathies but should be adapted to the pattern of disease within that particular family.

Heart transplantation Considered in patients with estimated 1y survival <50%.

Indications/contraindications Table 9.8.

Assessment Each eligible patient is assessed for psychosocial factors and physical factors (e.g. renal failure, obesity, age, peripheral vascular disease) which affect prognosis before a decision whether to place the patient on the transplant list is made.

Postoperatively Patients require lifelong immunosuppression—usually with ciclosporin. Follow-up is undertaken in specialist clinics.

Prognosis 1 in 4 patients die on the transplant list; 60% receive a transplant in <2y. Perioperative mortality is <10%; 1y survival 92%; 5y survival 75%; 10y survival 60%. Patients have accelerated graft atherosclerosis. Complications of immunosuppression include ↑ risk of infection and cancer.

Table 9.8 Indications and contraindications for heart transplant

Indications	Contraindications
All patients must have end stage heart disease. *Causes*:	Systemic disease likely to affect life expectancy (e.g. malignancy)
• IHD (50%)	Active infection (HIV, hepatitis B or C)
• Cardiomyopathy (40%)	Significant pulmonary vascular disease.
• Valvular and congenital heart defects (5%)	Continued excess alcohol consumption
	Significant cerebral/systemic vascular disease

Patient information and support

British Heart Foundation ☎ 0300 330 3311 🖱 www.bhf.org.uk
Cardiomyopathy Association ☎ 0800 018 1024 🖱 www.cardiomyopathy.org

Valve disease

Heart murmurs ➔ p. 206

⚠ All patients with newly detected valve disease, except those with mitral valve prolapse or aortic sclerosis, require cardiology referral.

- **Admit** If suspected endocarditis
- **Refer urgently/admit** If symptomatic valve disease or if valve disease underlies the presenting condition, e.g. heart failure caused by aortic stenosis, AF caused by mitral valve disease

Mitral stenosis Usually due to rheumatic fever.

Presentation

- **Symptoms** Breathlessness, palpitations, fatigue. May result in pulmonary hypertension which presents with right heart failure, haemoptysis and/or recurrent bronchitis
- **Signs** Peripheral cyanosis ('malar flush' on cheeks), left parasternal heave, tapping apex beat, AF, rumbling mid-diastolic murmur at the apex

Management Confirm with Echo. Refer to cardiology. Treatment is medical (treatment of AF, heart failure and anticoagulation) ± surgical (valvotomy, balloon valvoplasty, valve replacement).

Mitral regurgitation (incompetence) *Causes*:

- Congenital
- Rheumatic fever
- Mitral valve prolapse
- Ventricular dilatation
- Endocarditis
- Cardiomyopathy
- Ruptured papillary muscle/chordae tendineae following MI
- RA

Presentation

- **Symptoms** Dyspnoea, fatigue
- **Signs** Displaced apex (→ left axilla), pansystolic murmur at the apex radiating to axilla, AF, left ventricular failure

Management Confirm with Echo. Refer to cardiology. Treatment is medical (treatment of AF, heart failure and anticoagulation) ± surgical (valve replacement).

Mitral valve prolapse Prevalence ~1 in 20.

Presentation

- **Symptoms** Usually none. Rarely atypical chest pain, palpitations, syncope, postural hypotension, emboli
- **Signs** Late systolic murmur over apex

Management Confirm with Echo. If syncope or palpitations refer to cardiology—a rare complication is ventricular arrhythmia.

Aortic sclerosis Thickening and stiffening of the aortic valve not associated with outflow obstruction that occurs with age. Clinically an ejection systolic murmur is present but no other symptoms or signs. CXR may show a calcified valve. No treatment is required.

Aortic stenosis *Causes*:

- Congenital
- Rheumatic fever
- Bicuspid valve
- Degenerative calcification
- Hypertrophic cardiomyopathy

Presentation

- Symptoms Angina, breathlessness, syncope or 'funny turns', dizziness, sudden death
- Signs Small-volume pulse, low pulse pressure (difference between systolic and diastolic BP), ejection systolic murmur loudest in the aortic area which radiates to carotids and apex

Management Echo is diagnostic and gives an estimate of the gradient across the valve and thus severity of the condition. Refer to cardiology. Surgery (valve replacement or transcutaneous valvoplasty) is considered for those with syncope or if systolic gradient across the valve is >50mmHg. Avoid treatment with ACE inhibitors.

Aortic regurgitation *Causes*:

- Congenital, e.g. VSD
- Bicuspid aortic valve
- Rheumatic fever
- Aortic dissection
- Endocarditis
- Cardiomyopathy
- Syphilis
- Marfan's or Ehlers–Danlos syndrome

Presentation

- Symptoms Dyspnoea, palpitations (extrasystoles)
- Signs Prominent pulse ('water-hammer'), wide pulse pressure, visible neck pulsation (Corrigan's sign), head nodding in time with pulse (De Musset's sign), visible capillary pulsations (e.g. in nail bed—Quincke's sign), displaced apex beat, high-pitched early diastolic murmur (easily missed)

Management Confirm with Echo. Refer to cardiology for consideration of surgery.

Right heart valve disease Echo is diagnostic. Always requires specialist management.

- Tricuspid stenosis Mitral valve disease coexists. *Cause*: rheumatic fever. *Murmur*: early diastolic (left sternal edge in inspiration). Treatment is with diuretics ± surgery (valvotomy or replacement)
- Tricuspid regurgitation *Causes*: RV enlargement, endocarditis (IV drug misusers), carcinoid, rheumatic fever, congenital. Presents with oedema, breathlessness, pulsatile hepatomegaly (± jaundice), ascites, pansystolic murmur loudest at left sternal edge. Treatment is with diuretics, vasodilators, ± surgery (valve replacement or annuloplasty)
- Pulmonary stenosis *Causes*: congenital (Fallot's tetralogy), rheumatic, carcinoid. *Murmur*: ejection systolic murmur (loudest to left of upper sternum, radiating to left shoulder). ECG: RVH. *CXR*: dilated pulmonary artery. Treatment (if needed) is with pulmonary valvotomy
- Pulmonary regurgitation Due to pulmonary hypertension (➔ p. 236). *Murmur*: decrescendo early diastolic murmur at left sternal edge

⚠ Women planning pregnancy who have known valve disease require review for specialist advice.

Other structural abnormalities of the heart

Coarctation of the aorta Localized narrowing of the descending aorta usually distal to the origin of the left subclavian artery.

Presentation Heart failure, ↑ BP, murmur heard incidentally (ejection systolic murmur over the left side of the chest radiating to the back), lack of femoral pulses or radio-femoral delay. Rarely presentation is with a complication, e.g. subarachnoid haemorrhage or endocarditis. *CXR*—prominent left ventricle. *ECG*—Left ventricular hypertrophy.

Management Refer to cardiology—surgery to remove the narrowed portion of the aorta is usually indicated.

Atrial septal defect (ASD) A hole connects the 2 atria. Holes high in the septum (ostium secundum) are most common (2 in 1000 live births); holes lower in the septum (ostium primum) are associated with AV valve abnormalities. Blood flows from L → R through the shunt and the right heart takes the burden.

Presentation

- Ostium secundum defects Symptoms are rare in infancy and uncommon in childhood. If detected in these groups, presents as a murmur (systolic—loudest in the 2nd left interspace) found incidentally, with breathlessness or tiredness on exertion or recurrent chest infections. Presentation is usually in the 3rd or 4th decade with heart failure, pulmonary hypertension, and/or atrial arrhythmias
- Ostium primum defects Heart failure commonly develops in infancy/childhood ± severe pulmonary hypertension. In addition to the ASD murmur, there may be a pansystolic murmur signifying mitral or tricuspid valve regurgitation

Investigation

- CXR Cardiomegaly with a prominent right atrium ± pulmonary artery ± pulmonary plethora
- ECG Right axis deviation (ostium secundum defect) or left axis deviation (ostium primum defect), RVH ± RBBB
- Echo Diagnostic

Management Refer to cardiology. Cardiac surgery to close the defect is usually indicated.

Ventricular septal defect (VSD) A hole connects the 2 ventricles. Blood flows initially from L → R through the hole. May be congenital (2 in 1000 live births) or acquired (usually septal rupture post MI).

Acquired VSD Suspect if new pansystolic murmur ± heart failure develop after MI. Investigate as for congenital VSD. Refer to cardiology (speed of referral will depend on state of the patient) for advice on further management.

Congenital VSD

- Small VSD ('maladie de Roger') Normally asymptomatic. A thrill may be palpable at lower left sternal border; harsh pansystolic murmur—small holes give loud murmurs. CXR and ECG are normal. Diagnosis is confirmed on Echo. Refer to cardiology
- Moderate VSD Symptoms usually appear in infancy—breathlessness on feeding/crying, failure to thrive, recurrent chest infections. As the child gets older symptoms improve (relative size of the defect ↓). On examination, there may be cardiomegaly, a thrill palpable at the left sternal edge, and a pansystolic murmur. CXR shows cardiomegaly ± prominent pulmonary arteries ± pulmonary plethora. Diagnosis is confirmed on Echo. Refer to cardiology
- Large VSD Presents with heart failure at ~3mo of age though there may be symptoms of breathlessness on feeding/crying prior to then. On examination, the baby is obviously unwell—underweight, breathless, pulmonary oedema ± cyanosis, large heart, thrill over left sternal edge ± parasternal heave, murmur—often not pansystolic due to high right ventricular pressures. Admit to paediatrics—medical treatment ± surgery is always needed

Marfan's syndrome Autosomal dominant connective tissue disease causing abnormalities of fibrillin (a glycoprotein in elastic fibres). *Features include*:

- Arachnodactyly (long spidery fingers)
- High-arched palate
- Arm span > height
- Lens dislocation ± unstable iris
- Aortic dilatation (β-blockers appear to slow this)
- Aortic incompetence may occur, e.g. in pregnancy
- Aortic dissection may cause sudden death—Echo screening may be helpful for affected individuals

If suspected refer to cardiology ± genetics. There is currently no antenatal screening test available.

Other congenital heart disease ➔ p. 858

Aneurysms

An arterial aneurysm forms when there is a 50% ↑ in normal diameter of the vessel. Aneurysms may affect any medium/large artery—aorta/iliac arteries > popliteal > femoral > carotid. FH of aneurysm is a risk factor.

Causes Atheroma (most common); injury; infection (e.g. endocarditis, syphilis—mycotic aneurysms).

Abdominal aortic aneurysm (AAA) Prevalence is ~4% in men aged 65–74y (♂:♀ ~6:1). Acute rupture of AAA in the community has ~90% mortality accounting for 2% of deaths in ♂ aged >65y. Elective surgical repair has ~5–7% mortality.

Risk factors Smoking; ↑ BP; family history (risk ↑ ×4–10 if there is an affected 1st-degree relative).

Factors predisposing to rupture of AAA

- Diameter (risk ↑ with diameter)
- COPD
- Smoking
- ↑ diastolic BP
- FH
- Fast rate of expansion
- Inflammation within the aneurysm wall
- Thrombus free surface area of aneurysm sac

Presentation Often discovered as an incidental finding on abdominal examination, X-ray (calcification of aneurysm wall in 50% cases) or USS (75% asymptomatic at diagnosis). Otherwise presents with:

- **Local symptoms** Vague abdominal or back pain
- **Distant symptoms** Embolization/acute ischaemia of a limb. Multiple small infarcts (e.g. of toes) with good peripheral pulses suggests an aneurysm proximally
- **Collapse due to rupture** Hypovolaemic shock ± pulsatile abdominal mass ± abdominal or back pain—➔ p. 1060

Investigation USS confirms diagnosis, diameter, site, and extent.

Screening In the UK, all men aged 65y are offered aneurysm screening with a single abdominal USS. Men >65y can self-refer. Screening ↓ death from AAA by 44% over 4y. Possible screening results—Figure 9.4.

Management of abdominal aortic aneurysm

- **Acute rupture** ➔ p. 1060
- **Elective surgery** Refer if risk of rupture > risk elective repair. The greater the diameter, the more the risk (5.5cm diameter ≈10% 1y rupture rate; 10cm diameter >75% 1y rupture rate). AAAs >5.5cm are routinely repaired except if other factors ↑ risk of surgery; there is no survival benefit from treating smaller aneurysms. Refer urgently if symptomatic—may indicate rapid expansion, or inflammation—both risk factors for rupture
- **USS surveillance** Patients with AAAs <5.5cm diameter are screened at least annually. Routine repair takes place when and if the aneurysm expands to >5.5cm. 3 in 5 eventually warrant surgery

Inflammatory aneurysms Characterized by inflammatory infiltrate in the aneurysm wall. May be adherent to surrounding structures. *Presentation*: fever, malaise, and abdominal pain. Associated with ↑ mortality at operation.

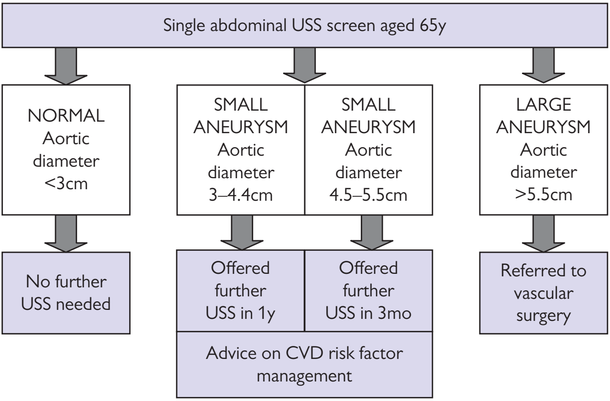

Single abdominal USS screen aged 65y
NORMAL Aortic diameter <3cm
SMALL ANEURYSM Aortic diameter 3–4.4cm
SMALL ANEURYSM Aortic diameter 4.5–5.5cm
LARGE ANEURYSM Aortic diameter >5.5cm
No further USS needed
Offered further USS in 1y
Offered further USS in 3mo
Referred to vascular surgery
Advice on CVD risk factor management

Figure 9.4 Possible AAA screening results

Thoracoabdominal aneurysm Involves thoracic and abdominal aorta—including the origins of the visceral and renal arteries. Surgery is more complex and carries higher mortality.

Dissecting thoracic aortic aneurysm ➔ p. 1060

Popliteal aneurysm 80% peripheral aneurysms. Most are >2cm diameter; 50% are bilateral. Associated with AAA (40%). Presents with acute below knee ischaemia secondary to aneurysm thrombosis or embolization. Popliteal pulses are pronounced. Diagnosis is confirmed on USS.

Management

- Acute ischaemia—➔ p. 1108
- Elective surgery (popliteal bypass)—when aneurysm >2.5cm diameter

Femoral artery aneurysm *Presentation* : local pressure symptoms, thrombosis, or distal embolization. *Surgical treatment*: bypass surgery.

Carotid artery aneurysm Rare. Presents with pulsatile lateral neck swelling ± carotid territory TIAs. Rarely can rupture. Refer to vascular surgery for surgical treatment.

Carotid body tumour Slow-growing tumour arising in the carotid body at the carotid bifurcation. Presents with a slowly enlarging mass which transmits carotid pulsation. Refer to vascular surgery for angiographic confirmation of diagnosis. Treatment is with surgical excision. If untreated, becomes locally invasive and may eventually metastasize.

Cerebral artery aneurysm ➔ p. 534

Further information

National Screening Aneurysm Screening Programme 🖱 www.gov.uk/topic/population-screening-programmes/abdominal-aortic-aneurysm

Patient information and support

British Heart Foundation ☎ 0300 330 3311 🖱 www.bhf.org.uk
Circulation Foundation 🖱 www.circulationfoundation.org.uk

Chronic peripheral ischaemia

Peripheral vascular disease (normally atherosclerotic) commonly affects arteries supplying the legs.

Prevalence 20% patients age >60y. Assess for the presence of peripheral arterial disease if:

- Symptoms suggestive of peripheral arterial disease *or*
- DM, non-healing wounds on the legs or feet or unexplained leg pain *or*
- Being considered for interventions to the leg or foot *or*
- Need to use compression hosiery

Natural history Most remain stable. A minority (20% over 10y) progress from intermittent claudication to critical limb ischaemia. Management of CVD risk factors is essential.

Intermittent claudication Restriction of blood flow causes pain on walking. *Risk factors*:

- ♂ > ♀
- Smoking
- Obesity
- ↑ BP
- Hyperlipidaemia
- DM
- Physical inactivity
- Hypercoagulable states
- Postmenopausal

Presentation Presents with muscular, cramp-like pain in the calf, thigh or buttock on walking that is rapidly relieved on resting. The leg is cool and white with atrophic skin changes and absent pulses (Table 9.9):

- Disease in the superficial femoral artery Absent popliteal and foot pulses. Causes calf claudication
- Disease of the aorta or iliac artery Weak or absent femoral pulse ± femoral bruit. Causes calf, thigh, or buttock claudication

Differential diagnosis Nerve root compression, e.g. sciatica; spinal stenosis—usually bilateral pain which may occur after prolonged standing as well as exercise—not rapidly relieved by rest.

Investigation

- Blood FBC, U&E, Cr, eGFR (peripheral vascular disease is associated with renal artery stenosis—➔ p. 417), HbA1c, lipids
- Ankle–brachial systolic pressure index (ABPI) Good history + ABPI <0.95 confirms diagnosis; ❗ do not exclude peripheral arterial disease in people with DM based on normal or ↑ ABPI
- Duplex USS Used to determine site of disease (may only be available via secondary care referral)

Management

- Exercise Offer a supervised programme (ideally 2h/wk for 3mo) to all patients; encourage to exercise to the point of maximal pain
- ↓ risk factors Patients with claudication have a 3× ↑ risk of death from MI/stroke. Advise to stop smoking, moderate alcohol, and lose weight. Ensure optimum treatment of ↑ BP, lipids, and DM
- Antiplatelet agents Treat all patients with aspirin 75mg od (or clopidogrel 75mg od if aspirin-intolerant)
- Foot care Regular chiropody

Referral to vascular surgery

E = Emergency admission; *U* = Urgent; *S* = Soon; *R* = Routine

- Critical limb ischaemia—*E/U*
- Severe symptoms—*S*
- Uncertainty about diagnosis—*R*
- No better after exercise training—*R*

Table 9.9 Location of the pulses of the lower limbs

Pulse	Location
Femoral	Below inguinal ligament; ⅓ of the way up from pubic tubercle
Popliteal	With knee flexed at right angles palpate deep in the midline
Posterior tibial	1cm behind medial malleolus
Dorsalis pedis	Variable—on the dorsum of the foot just lateral to the tendons to the big toe. ❗ Many healthy people have only 1 foot pulse

Critical limb ischaemia

Presentation Deteriorating claudication and nocturnal rest pain (usually just after fallen asleep—hanging the foot out of bed improves the pain). Ulceration or gangrene results from minor trauma.

Examination Look for:

- Atrophic skin changes—pallor, cool to the touch, hairless, shiny
- On lowering the leg turns a dusky blue-red colour; on elevation—pallor and venous guttering
- Ulceration—check under the heel and between the toes
- Swelling suggests the patient is sleeping in a chair to avoid rest pain or, rarely, pain from deep infection
- Absent foot pulses—if present consider alternative diagnosis
- ABPI <0.5—❗ arterial calcification can result in falsely high readings

Management Analgesia (often requires opioid); refer for urgent vascular surgical assessment.

The diabetic foot ➜ p. 330

Specialist management of peripheral arterial disease

- **Angiography** to assess extent and position of disease
- **Percutaneous transluminal angioplasty ± stenting** Most suitable for short occlusions/stenoses of the iliac and superficial femoral vessels. 1y patency rate 80–90%
- **Surgery** Most suitable for longer occlusions/multiple stenoses—aortobifemoral bypass grafts have 5y patency rates >90%; femoropopliteal bypass grafting gives 5y patency rates of <70%. Aspirin ↓ risk of re-occlusion. Amputation is a last option

Drug treatment of intermittent claudication Naftidrofuryl ↑ walking distance but it is unclear whether it influences outcome. Consider only if supervised exercise has not led to improvement and the person does not want or is unsuitable for angioplasty or bypass surgery. Reassess after 3–6mo. Discontinue if no improvement.

Acute limb ischaemia ➜ p. 1108

Further information

NICE (2012, updated 2018) Lower limb peripheral arterial disease: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg147

Patient information and support

Circulation Foundation 🖱 www.circulationfoundation.org.uk

Varicose veins

Tortuous, twisted, or lengthened veins. *Prevalence*: 17–31%. ♂ > ♀ (≈5:4). The vein wall is inherently weak leading to dilatation and separation of the valve cusps so they become incompetent. Blood flows backwards from the deep to superficial venous system, causing back pressure and further dilatation.

Most varicose veins are primary. *Risk factors*: age, parity, occupations requiring a lot of standing, obesity (women only). 2° *causes*: DVT, pelvic tumour, pregnancy, or AV fistula.

Types

- **Trunk** Varicosities of the long or short saphenous vein or their branches. May be symptomatic
- **Reticular** Usually asymptomatic. Dilated tortuous subcutaneous veins not belonging to the main branches of the long or short saphenous vein
- **Telangiectasia** Intradermal venules <1mm—spider veins, thread veins, star bursts, matted veins. Unsightly but otherwise asymptomatic

Presentation Consider:

- **Why is the patient consulting now?** Patients are often worried about appearance of varicose veins or prognosis if left untreated but have no other symptoms (1 in 3 consultations)
- **Symptoms** Heaviness, tension, aching (worse on standing and in the evening; improved by elevating the leg and support stockings), itching
- **Complications**
- **PMH** Previous surgery or injection for varicose veins; pregnancy; past history of DVT or thrombophlebitis; CHC or HRT
- **FH** Varicose veins or DVT

Examination

- **Abdominal examination** To exclude secondary causes
- **Veins** With the patient standing, inspect distribution of the veins and any secondary skin changes. Patterns of distribution:
  - *Long saphenous distribution*: thigh and medial aspect of the calf
  - *Short saphenous distribution*: below the knee on the posterior and lateral aspects of the calf

Management Reassurance is often all that is needed.

- **If symptoms are troublesome** Advise support stockings; avoid standing for prolonged periods and if standing do not stand still; walk regularly; ↓ weight (if obese)
- **If any complications or severe symptoms** Refer for vascular surgical assessment. In general, patients with purely cosmetic problems are not treated under the NHS

⚠ Check ABPI to exclude significant arterial disease before recommending compression hosiery (ABPI should be >0.8).

Bleeding varicose veins Bleeding can be stemmed by raising the foot above the level of the heart and applying compression. If the patient is fit for surgery, refer for surgical assessment. Once recovered from the bleed, advise compression hosiery if ABPI >0.8.

Complications

- Haemorrhage
- Varicose eczema
- Skin pigmentation
- Thrombophlebitis
- Lipodermatosclerosis—fibrosis of the dermis and subcutis around the ankle resulting in firm induration
- Oedema
- Venous ulceration—40% do not have visible varicose veins.
- Atrophie blanche—white, lacy scars

CHC and HRT Women with varicose veins taking CHC or HRT are not at ↑ risk of DVT but are at ↑ risk of thrombophlebitis.

Saphena varix Dilatation of the saphenous vein at its confluence with the femoral vein which transmits a cough impulse. May have bluish tinge and disappears on lying down. A cause of a lump in the groin. Action only needed if symptomatic.

Thrombophlebitis Presents as severe pain, erythema, pigmentation over, and hardening of the vein. Thrombophlebitis in varicose veins results from stasis. Consider underlying malignancy or thrombophilia if thrombophlebitis occurs in normal veins or there is recurrent thrombophlebitis in varicose veins.

Management ❗ There is no indication for antibiotics.

- Crepe bandaging to compress vein and minimize propagation of thrombus (if ABPI >0.8)
- Analgesia—preferably NSAID
- Ice packs and elevation
- Low-dose aspirin—75–150mg od

> ⚠ If phlebitis extends up the long saphenous vein towards the saphenofemoral junction, refer for urgent duplex scanning—saphenofemoral ligation may be indicated if thrombus extends into the femoral vein.

Follow-up If the patient is fit for surgery, refer for surgical assessment as thrombophlebitis tends to recur if the underlying venous abnormality is not corrected.

History of thrombophlebitis is a relative contraindication to CHC (➔ p. 731). Evidence regarding HRT is less clear.

Thrombophlebitis migrans Recurrent tender nodules affecting veins throughout the body. Associated with carcinoma of the pancreas.

Patient information

Circulation Foundation 🖱 www.circulationfoundation.org.uk

Deep vein thrombosis

DVT may be proximal—involving veins above the knee—or isolated to the calf veins. It may also occur in the cerebral sinus, and veins of the arms, retina, and mesentery. *Incidence*: 1 in 1000 people/y. *Risk factors*:

- Age >40y
- Smoking
- Obesity
- Immobility
- Recent long-distance travel
- Pregnancy
- Puerperium
- CHC/HRT use
- Surgery
- Recent trauma
- Malignancy
- Heart failure
- Nephrotic syndrome
- Inflammatory bowel disease
- PMH of venous thromboembolism
- Inherited thrombophilic clotting disorders
- Other chronic illness

❗ Central venous catheters are a common cause of upper limb DVT.

Presentation Unilateral leg pain, swelling, and/or tenderness ± mild fever, pitting oedema, warmth, and distended collateral superficial veins.

Differential diagnosis

- Cellulitis
- Arthritis/muscle tear
- Ruptured Baker's cyst
- Superficial thrombophlebitis
- Chronic venous insufficiency
- Venous obstruction
- Post-thrombotic syndrome
- Acute arterial ischaemia
- Lymphoedema
- Fracture
- Hypoproteinaemia

Immediate action Clinical diagnosis is unreliable. <50% with clinically suspected DVT have diagnosis confirmed on diagnostic imaging. In most areas in the UK, rapid access DVT assessment clinics operate.

⚠ If there will be a delay in investigation to exclude DVT, provide anticoagulation with a direct oral anticoagulant (DOAC) or low-molecular-weight heparin (LMWH) in the interim.

Clinical prediction rules (e.g. Wells' score—Box 9.2) are used to decide whether patients fall into high or low probability groups for DVT.

- **If low probability** Do a blood D-dimer. If −ve, DVT is excluded. If +ve, assess the patient as if medium/high probability
- **If medium/high probability** Compression USS assessment is undertaken ± D-dimer. If USS is negative and low probability or −ve D-dimer, DVT is excluded. If USS is +ve, diagnosis of DVT is confirmed. If USS is −ve and medium/high probability or +ve D-dimer, USS is repeated after 1wk or the patient is assessed with venography, CT, or MRI

D-dimer testing Detects a degradation product of fresh venous thrombus. It may be available as a near-patient test with a result in <15min in some practices—do not delay referral to await result if near-patient testing is not available. A normal D-dimer result has a high negative predictive value making DVT unlikely. However, raised D-dimer levels are not specific for venous thromboembolism. Other causes of ↑ D-dimer include:

- Malignancy
- Pregnancy
- Wound healing
- Recent trauma
- Inflammation
- Anticoagulant use
- Sepsis
- Liver impairment

Management of patients with confirmed DVT

- Initial anticoagulation is as an outpatient with either a direct oral anticoagulant (e.g. rivaroxaban, apixaban) *or* LMWH followed by oral anticoagulation (usually warfarin). If anticoagulating with warfarin, LMWH should be continued for at least 4d and until INR is in therapeutic range (target INR 2.5; range 2–3) for ≤2d
- Oral anticoagulants ↓ risk of further thromboembolism and should be continued for 3–6mo after a single DVT (➔ p. 648)
- Graduated elastic compression stockings—should be worn for >2y as they ↓ risk post-thrombotic leg syndrome by 12–50%

❗ If a patient has a DVT and there is no obvious cause: if <45y, consider thrombophilia; if >45y, consider undiagnosed cancer.

Management during pregnancy ➔ p. 802

Complications of DVT

- Pulmonary embolus Without treatment 20% with proximal DVT develop PE (➔ p. 1070)
- Post-thrombotic syndrome Occurs after DVT. Results in chronic venous hypertension causing limb pain, swelling, hyperpigmentation, dermatitis, ulcers, venous gangrene, and lipodermatosclerosis
- Recurrent venous thromboembolism Patients with history of DVT or PE have ↑ risk of recurrence in high-risk situations (trauma, surgery, immobility, pregnancy) and should receive prophylaxis with heparin/oral anticoagulants in such situations

Box 9.2 Wells' diagnostic algorithm

Score 1 point if:

- Active cancer (ongoing treatment or treatment in the past 6mo, or palliative care)
- Paralysis, paresis, or recent plaster immobilization of the legs
- Recently bedridden for ≥3d, or major surgery in the past 12wk (GA or regional anaesthesia)
- Localized tenderness along the distribution of the deep vein system (e.g. back of the calf)
- Entire leg swelling
- Calf diameter of affected leg (measured 10cm below the tibial tuberosity) >3cm greater than that of the unaffected leg
- Pitting oedema of affected but not unaffected leg
- Collateral superficial veins (non-varicose)
- Previous DVT

Take away 2 points if:

An alternative cause is as/more likely than DVT.

Interpretation

- If score is <2—DVT is unlikely
- If DVT is ≥2—DVT is likely

Source: data from Wells PS *et al.*, Value of assessment of pretest probability of deep-vein thrombosis in clinical management, *The Lancet*, **350**, 1795–8.

Further information

NICE (2012, updated 2015) Venous thromboembolic diseases. ☞ www.nice.org.uk/guidance/cg144

Chapter 10

Respiratory medicine

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Breathlessness

Dyspnoea Sensation of shortness of breath. Speed of onset helps diagnosis (Table 10.1). Try to quantify exercise tolerance (e.g. dressing, distance walked, climbing stairs).

Acute breathlessness ➔ p. 1068

Exertional dyspnoea Breathlessness with exercise. Causes are the same as dyspnoea generally. The New York Heart Association classifies the severity of functional limitations in patients with heart failure:

- Class 1 Normal. No limitations
- Class 2 Slight limitation during ordinary activity
- Class 3 Marked limitation during less than ordinary activity, e.g. walking short distances
- Class 4 Inability to do any activity. Symptoms present at rest

Orthopnoea Dyspnoea on lying flat and relieved by sitting up. Associated with left heart dysfunction, e.g. LVF.

Paroxysmal nocturnal dyspnoea Acute form of dyspnoea that causes the patient to awake from sleep. The patient is forced to sit upright or stand out of bed for relief. Associated with pulmonary oedema.

Combined chest pain and dyspnoea Consider:

- MI
- Pericarditis
- Dissecting aneurysm
- PE
- Oesophageal pain
- Musculoskeletal pain
- Chest infection
- Pulmonary malignancy

⚠ Refer any patient with symptoms/signs of superior vena cava obstruction (acute breathlessness, headache worse on stooping, swelling of the face and/or neck, with fixed elevation of jugular venous pressure) for immediate medical or oncology assessment[N].

Offer urgent CXR if ≥40y and ≥2 (or if smoker/ex-smoker or history of asbestos exposure and ≥1) of the following unexplained symptoms: cough; fatigue; shortness of breath; chest pain; weight ↓; appetite ↓[N].

Respiratory rate Normal values vary according to age:

- <1y: 30–40 breaths/min
- 1–2y: 25–35 breaths/min
- 2–5y: 25–30 breaths/min
- 5–12y: 20–25 breaths/min
- >12y: 15–20 breaths/min

↑ *respiratory rate* Consider:

- Lung disease, e.g. pneumonia, asthma
- Heart disease, e.g. LVF
- Metabolic disease, e.g. ketoacidosis
- Drugs, e.g. salicylate overdose
- Psychiatric causes, e.g. hyperventilation

↓ *respiratory rate* Consider:

- CNS disease, e.g. CVA
- Drugs, e.g. opioids

Pneumothorax ➔ p. 1070

Hyperventilation May be fast (>20 breaths/min) or deep (tidal volume ↑). If inappropriate, results in palpitations, dizziness, faintness, tinnitus, chest pains, perioral and peripheral tingling (due to plasma Ca^{2+} ↓).

Table 10.1 Causes of dyspnoea

Cause	Acute	Subacute	Chronic
Cardiac disease	Acute LVF Arrhythmia Acute MI Aortic dissection Tamponade	Arrhythmia Subacute bacterial endocarditis Pericarditis	CCF Valvular disease, e.g. mitral stenosis Congenital heart disease
Lung disease	Acute asthma attack COPD exacerbation Upper airway obstruction Pneumonia Acute pneumonitis, e.g. due to inhaling toxic gas Pulmonary embolus Pneumothorax	Asthma COPD exacerbation Pneumonia Pleural effusion Lobar collapse	Asthma COPD Cystic fibrosis Interstitial lung disease Occupational lung disease Mesothelioma Lung cancer
Other	Hyperventilation Foreign body inhalation Guillain–Barré syndrome Altitude sickness Ketoacidosis Polio Musculoskeletal pain Oesophageal pain	Aspirin poisoning Myasthenia gravis Thyrotoxicosis Superior vena cava obstruction	Kyphoscoliosis Obesity Anaemia Neuromuscular weakness, e.g. MND, MS

Causes include:

- Anxiety (most common cause)
- Early pulmonary oedema
- PE
- Hyperthyroidism
- Fever
- Lymphangitis
- Weakness of the respiratory muscles

Kussmaul respiration Deep, sighing breathing that is principally seen in metabolic acidosis, e.g, diabetic ketoacidosis and uraemia.

Neurogenic hyperventilation Stroke, tumour, or CNS infection.

Hypoventilation Abnormally ↓ pulmonary ventilation. Respiration may be too slow or tidal volume ↓. *Causes include*:

- Respiratory depression, e.g. opioid analgesia, anoxia, trauma
- Neurological disease, e.g. Guillain–Barré disease; polio; motor neurone disease; syringobulbia
- Lung disease, e.g. pneumonia, collapse, pneumothorax, pleural effusion
- Respiratory muscle disease, e.g. myasthenia gravis, dermatomyositis
- Limited chest movement, e.g. kyphoscoliosis

Cheyne–Stokes respiration Breathing becomes progressively deeper and then shallower (± episodic apnoea) in cycles. *Causes*: brainstem lesions/compression (stroke, ↑ ICP); chronic pulmonary oedema; poor cardiac output. It is enhanced by narcotics.

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. www.nice.org.uk/guidance/ng12

Cough

A cough is a reaction to irritation anywhere from pharynx to lungs.

Acute cough (<3wk) *Causes*:

- URTI
- Croup
- Tracheitis
- Acute bronchitis
- Pneumonia—productive, loose cough
- Acute exacerbation of normally well-controlled asthma
- Inhaled foreign body—especially in well children

Reserve CXR for patients with marked focal chest signs or where inhalation of foreign body or lung cancer is suspected.

Management Treat the cause where possible; steam inhalation often eases symptoms temporarily; review if not clearing.

Reasons to prescribe antibiotics immediately[N] Investigate further and/or give antibiotics (e.g. amoxicillin 500mg tds/clarithromycin 500mg bd/doxycycline 100mg od) immediately if the patient:

- Is systemically very unwell or has symptoms/signs suggestive of serious illness and/or complications, e.g. pneumonia
- Is at high risk of serious complications because of pre-existing co-morbidity, e.g. significant heart, lung, renal, liver, or neuromuscular disease, immunosuppression, CF, and young children born prematurely
- Is aged >65y with acute cough and ≥2 or more of the following, or aged >80y with acute cough and ≥1 of the following:
    - Hospitalization in the previous year
    - Type 1 or type 2 DM
    - History of congestive heart failure
    - Current use of oral glucocorticoids

Chronic cough (>3wk) *Causes*:

- Postnasal drip
- Post viral
- COPD/asthma
- Lung cancer
- Pertussis
- TB
- Bronchiectasis
- Pulmonary oedema
- Foreign body
- Vocal cord palsy
- GORD
- LVF
- Drug induced (e.g. ACE inhibitors)
- Smoker's cough
- Ear wax
- Psychogenic
- Idiopathic

⚠ *Red flags:* Weight ↓, night sweats, fever, haemoptysis.

Management Offer an urgent CXR in those ≥40y if they have ≥2 (or if smoker/ex-smoker or exposed to asbestos and ≥1) of the following symptoms: cough; fatigue; shortness of breath; chest pain; weight ↓; appetite ↓[N]. Treat the cause. If no cause is found, refer.

Sputum ❗ Absolutely clear sputum is probably saliva.

- Smoking is the leading cause of excess sputum production—look for black specks of inhaled carbon
- Yellow-green sputum is due to cell debris (bronchial epithelium, neutrophils, eosinophils) and is not always infected
- Bronchiectasis causes copious greenish sputum
- Blood-stained sputum (haemoptysis) always needs full investigation
- Pink froth suggests pulmonary oedema

Haemoptysis Expectoration of blood/blood-stained sputum. *Causes*:

- Infection—bronchitis, pneumonia, lung abscess, TB
- Violent coughing
- Bronchiectasis
- Lung cancer
- PE (blood is not mixed with sputum)
- Inhaled foreign body
- Iatrogenic: anticoagulation, endotracheal tube
- Trauma
- Cardiac: acute LVF, mitral stenosis
- Blood dyscrasia/bleeding diathesis
- Idiopathic pulmonary haemosiderosis
- Bronchial adenoma
- Mycosis, e.g. aspergilloma
- Goodpasture's syndrome
- Collagen vascular disease, e.g. PAN, granulomatosis with polyangiitis
- Idiopathic

❗ Differentiate from haematemesis or local bleeding from the nasopharynx or sinuses. Melaena may occur if enough blood is swallowed.

Management *Always* requires investigation to find the cause.

- Admit as an acute medical emergency if the patient is compromised by the bleeding (i.e. tachycardia, low BP, postural drop) or has symptoms/signs of a cause requiring acute admission (e.g. PE, acute LVF)
- Refer for urgent chest physician assessment if aged ≥40y with unexplained haemoptysis[N]

❗ In patients with lung cancer who have a massive haemoptysis as a terminal event, consider treating with IV morphine/diamorphine and a sedative (e.g. midazolam or rectal diazepam) rather than admitting.

Bronchiectasis Consider in patients with persistent or recurrent chest infections. Permanently dilated bronchi act as sumps for infected mucus. *Causes*:

- Congenital CF, Kartagener syndrome
- Post-infection TB, pertussis, measles, pneumonia
- Other Bronchial obstruction, aspergillosis (➔ p. 299), hypogammaglobulinaemia (➔ p. 658), gastric aspiration

Presentation

- Mild cases Usually asymptomatic with winter exacerbations consisting of fever, cough, purulent sputum, pleuritic chest pain, dyspnoea
- More severe cases Persistent cough and sputum, haemoptysis, clubbing, low-pitched inspiratory and expiratory crackles and wheeze

Investigations CXR; sputum—M,C&S; spirometry—reversible airways obstruction is common; high-resolution CT detects disease in 97% of cases.

Management Refer to a respiratory physician. Treatment includes physiotherapy, antibiotics, bronchodilators, vaccination (influenza and pneumococcal) and (rarely) surgery.

Further information

NICE (2008) Respiratory tract infections: antibiotic prescribing. 🖱 www.nice.org.uk/guidance/cg69

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Chest signs

Signs associated with common chest pathology Table 10.2

Chest deformity

- **Barrel chest** The anteroposterior diameter of the chest is high compared to the lateral diameter, and expansion is ↓. Ribs move in a pump handle, up-and-down motion. Associated with chronic hyperinflation (e.g. asthma or COPD)
- **Pigeon chest (pectus carinatum)** Prominent sternum and flat chest associated with history of chronic childhood asthma or rickets
- **Funnel chest (pectus excavatum)** The lower end of sternum is depressed. Often inherited or idiopathic and usually harmless
- **Kyphosis** ↑ forward spinal convexity usually affecting thoracic spine:
  - *Postural* ('drooping shoulders' or 'roundback'): is common and voluntarily correctable
  - *Structural*: cannot correct voluntarily. *Causes:* osteoporosis, Paget's disease, ankylosing spondylitis, Scheuermann's disease. May cause a restrictive ventilatory defect and eventually respiratory failure
- **Scoliosis** ➔ p. 453
- **Harrison's sulcus** Groove deformity of the lower ribs at the diaphragm attachment site. Suggests chronic childhood asthma or rickets
- **Scars** Are there any scars indicative of previous chest surgery?

Chest expansion Expansion should be symmetrical and equal. If not, suspect chest pathology (e.g. consolidation, collapse, pneumothorax, effusion) on the side with ↓ movement.

Vocal fremitus or resonance

- **↑ transmission** implies consolidation. Even whispered sounds are heard clearly with a stethoscope (*whispering pectoriloquy*)
- **↓ transmission** implies something in the way blocking the transmission of sound. *Consider*: air (e.g. pneumothorax), fluid (e.g. effusion), pleural thickening (e.g. mesothelioma)

Percussion Define any areas of dullness to percussion by percussing from a resonant to dull area. *Interpretation*:

- **↑ resonance**—emphysema or pneumothorax (➔ p. 1070)
- **↓ resonance**—consolidation, collapse, abscess, tumour, fibrosis
- **Stony dullness**—pleural effusion

Breath sounds Assess character of breath sounds and added sounds:

- **Bronchial breathing** Breath sounds are harsher than normal and there is an audible gap between inspiration and expiration—often caused by lung consolidation, e.g. due to pneumonia
- **↓ breath sounds** Consider: pleural effusion, pneumothorax, emphysema, lung collapse
- **Added sounds** Pleural rub; wheeze; crepitations/crackles

Wheeze Musical sound heard during expiration.

- **Polyphonic** Narrowing of many small airways; typical of asthma/COPD
- **Monophonic** Indicates single large airway obstruction, e.g. due to foreign body or tumour

Crackles in the chest Produced by air flow moving secretions.

- **Fine crackles** Consider pulmonary oedema (early inspiratory—usually best heard at the lung bases at the back); early pneumonia; fibrosing alveolitis (late inspiratory)
- **Coarse crackles** Consider TB; resolving pneumonia; bronchiectasis; lung abscess

Pleural rub Creaking sound produced by movement of visceral over parietal pleura when both are inflamed (e.g. pneumonia, infarction).

Pleural effusion Fluid in the pleural cavity. Simple effusions may be transudates (<30g/L protein) or exudates (>30g/L protein). Effusions may also be blood, lymph, or pus (empyema). *Causes of simple effusion*:

- Malignancy, e.g. lung cancer, mesothelioma, Meig's syndrome
- Infection, e.g. pneumonia, TB
- Infarction (pulmonary embolus)
- Heart failure
- Constrictive pericarditis
- Inflammation, e.g. SLE, RA, pancreatitis, asbestos exposure
- Hypoproteinaemia
- Hypothyroidism

Presentation May be incidental finding on CXR. *Symptoms*: dyspnoea, pleuritic pain, symptoms of underlying cause. *Signs*: absent breath sounds, dullness to percussion, ↓ tactile vocal fremitus, ↓ vocal resonance. Above the effusion there is usually a zone of bronchial breathing. Early on there may be a pleural rub. Large effusions shift the mediastinum away from the affected side and there may be ↓ chest wall movement. Confirm with CXR. If cause is not apparent, refer for diagnostic tap.

Management Treat the underlying cause. Refer for drainage if symptomatic. Repeated drainage ± pleurodesis may be necessary.

Surgical emphysema Air in the subcutaneous tissue. Can be caused by spontaneous pneumothorax or trauma to the chest wall. Tissues appear swollen and crackle on palpation

Table 10.2 Chest signs associated with common chest pathology

	Consolidation, e.g. pneumonia	Pleural effusion	Collapsed lung	Pneumothorax
Mediastinum	Not displaced	Normal or displaced away from the effusion	Displaced towards the side of collapse	Displaced away from the side of pneumothorax
Expansion	↓	↓	↓	↓
Percussion	Dull	Stony dull	Dull	Hyper-resonant
Breath sounds	Bronchial breathing	↓	↓	↓
Added sounds	Crackles ± rub	Bronchial breathing above effusion	None	None
Other	↑ vocal resonance, whispering pectoriloquy	↓ vocal resonance		↓ vocal resonance

Other signs of respiratory disease

The trachea

- Palpate the trachea in the supraclavicular notch in the midline
- Deviation to the left or right suggests a shift of the upper mediastinum to that side
- The distance between the suprasternal notch and cricoid cartilage in an adult is 2–3 finger breadths. If it is less than this, the lungs are probably hyperinflated

Weight loss Non-specific symptom or sign. *Consider*:

- **GI causes** Malabsorption, malnutrition, dieting
- **Chronic disease** Hyperthyroidism, DM, COPD, heart failure, renal disease, degenerative neurological/muscle disease, chronic infection (e.g. TB, HIV)
- **Malignancy**
- **Psychiatric causes** Depression, dementia, anorexia

⚠ *Offer an urgent CXR*^{N} to any patient aged ≥40y with unexplained weight ↓ if:

- Ever smoked or exposed to asbestos
- Never smoked but with any of the following: cough or fatigue or shortness of breath or chest pain or appetite loss

Cachexia Severe generalized muscle wasting. *Causes*: neoplasia; malnutrition; chronic infection (e.g. TB); prolonged inactivity; dementia.

Night sweats *Consider*: TB; lymphoma; leukaemia; solid tumour (e.g. renal carcinoma); menopause; anxiety states.

Erythema nodosum ➔ p. 568

Peripheral oedema ➔ p. 204

Horner's syndrome Sympathetic nerve disruption to the iris causes:

- Small (meiotic) pupil with lack of pupil dilation in the dark
- Partial lid ptosis
- Anhidrosis of the forehead ±
- Enophthalmos

Causes

- Pancoast, cervical cord, or mediastinal tumour
- Aortic aneurysm
- Posterior inferior artery or basilar artery occlusion
- Hypothalamic lesion
- Syringomyelia

Pallor Check eyes/mucous membranes for pallor suggesting anaemia.

Cyanosis ➔ p. 204

Persistent thrombocytosis ➔ p. 636

Flapping tremor/asterixis Bilateral motor disturbance. Ask the patient to hold his hands straight out in front of him and dorsiflex his hands—this provokes a flapping, asynchronous tremor which is absent at rest. Due to CO_2 retention in severe COPD.

Lymphadenopathy ➔ p. 916

⚠ Consider an urgent CXR in any patient ≥40y with supraclavicular lymphadenopathy or persistent cervical lymphadenopathy[N].

⚠ Consider an urgent referral using a suspected cancer pathway if there is an unexplained neck lump in a person ≥45y, or there is a persistent unexplained neck lump in a person <45y[N].

Clubbing ➔ p. 577

⚠ Consider an urgent CXR to assess for lung cancer in people ≥40y with finger clubbing[N].

Yellow nails ➔ p. 576

Hoarseness ➔ p. 914

⚠ Consider a suspected cancer pathway referral for laryngeal cancer in people ≥45y with persistent unexplained hoarseness.

Stridor ➔ p. 915

Jugular venous pressure ➔ p. 205

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral.
🖱 www.nice.org.uk/guidance/ng12

Respiratory investigations

Indications for urgent CXR^{N} If ≥40y and:
- ≥2 (or if previously smoked or been exposed to asbestos ≥1) of the following unexplained symptoms: cough; fatigue; shortness of breath; chest pain; weight ↓; appetite ↓
- Any of the following Persistence of recurrent chest infection; finger clubbing; supraclavicular or persistent cervical lymphadenopathy; chest signs consistent with lung cancer or pleural disease; or thrombocytosis

Incidental findings on CXR/CT scan of emphysema/signs of chronic airways disease. Perform respiratory review and spirometry. Treat according to results. If no symptoms + normal spirometry:
- If current smoker Offer smoking cessation advice (➲ p. 156); ↑ risk of COPD/lung cancer; advise to return if new symptoms
- If non-smoker Ask about FH of lung/liver disease—consider α1-antitrypsin deficiency (➲ p. 398); advise to return if new symptoms

Peak flow Simple and cheap test. Poor measure of airflow limitation; tends to overestimate lung function. Best used to monitor progress of disease and effects of treatment for patients with asthma. Link with self-management plan (➲ p. 281). Meters are available on NHS prescription—EN 23747 (2007)/EU standard peak flow meters are supplied. Charts are available from NHS supplies (Form FP1010) and drug companies.

Measuring peak expiratory flow rate (PEFR) Normal values—Table 10.4, (➲ p. 274). Ask the patient to stand up (if possible) and hold the meter horizontally. Check the indicator is at 0 and the track is clear:
- Ask the patient to take a deep breath and blow out forcefully into the peak flow meter ensuring lips are sealed firmly around the mouthpiece
- Read the PEFR off the meter. The best of 3 attempts is recorded
- Consider using a low-range meter if predicted/best PEFR is <250L/min

Spirometry Measures the volume of air the patient (adult or child >5y) is able to expel from the lungs after a maximal inspiration.
- FEV$_1$ Volume of air the patient is able to exhale in the first second of forced expiration
- FVC Total volume of air the patient can forcibly exhale in 1 breath
- FEV$_1$/FVC Ratio of FEV$_1$ to FVC expressed as a %

Measuring FEV$_1$ and FVC Interpreting results—Table 10.3; normal values—Table 10.5, ➲ p. 275. Sit the patient comfortably and ask the patient to take a deep breath in:
- Ask the patient to blow the whole breath out as hard as possible until there is no breath left to expel and ensuring lips are sealed firmly around the mouthpiece. Encourage the patient to keep breathing out
- Repeat the procedure × 2 (i.e. 3 attempts in all); ≥2 readings should be within 100mL or 5% of each other

Flow volume measurement Figure 10.1

❗ In England, staff providing/interpreting spirometry require certification by March 2021. Competencies will be assessed every 3y.

RCP 3 questions Useful tool to identify patients with poor asthma control in general practice and monitor effect of changes of treatment. Morbidity categories correlate with lung function.

In the last month

- Have you had any difficulty sleeping because of your asthma symptoms (including cough)?
- Have you had your usual asthma symptoms during the day (cough, wheeze, chest tightness, or breathlessness)?
- Has your asthma interfered with your usual activities, e.g. housework, work/school, etc.?

NO to all questions = low morbidity
1 × YES answer = medium morbidity
2 or 3 × YES answer = high morbidity

❗ Alternatives include the Asthma Control Questionnaire (ACQ) and Asthma Control Test (ACT)/Children's Asthma Control Test. These questionnaires are not designed for use during an acute attack.

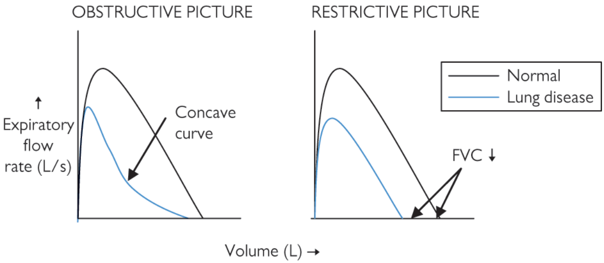


Figure 10.1 Flow–volume curves for patients with restrictive and obstructive lung disease

Reproduced from p.15 of the British Thoracic Society Guidelines Spirometry in Practice, with permission from the British Thoracic Society. www.brit-thoracic.org.uk

Table 10.3 Interpretation of spirometry results

	Restrictive lung disease, e.g. interstitial lung disease	Obstructive lung disease, e.g. COPD
FEV$_1$ *(% of predicted normal)*	↓ (<80%)	↓ (<80%)
FVC *(% of predicted normal)*	↓ (<80%)	Normal or ↓
FEV$_1$*/FVC*	Normal (>70%)	↓ (<70%)

Further information

ARTP/BTS Certificate in spirometry. 🖱 www.artp.org.uk
British Thoracic Society (BTS) (2013) A guide to performing quality assured diagnostic spirometry. 🖱 https://www.brit-thoracic.org.uk/media/70454/spirometry_e-guide_2013.pdf
NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Table 10.4 Predicted PEFR measurements in L/min (EU scale)

Children Height is the only determinant of PEFR in children. With ↑ age the pattern of adult values takes over.

Height: Feet	3′	3′4″	3′8″	4′	4′4″	4′8″	5′	5′4″	5′8″	6′
Metres	90cm	1	1.1	1.2	1.3	1.4	1.5	1.6	1.7	1.8
PEFR L/min	88	105	136	172	220	265	313	371	427	487

Women

Height: →

Feet	4′10″	4′11″	5′	5′1″	5′2″	5′3″	5′4″	5′5″	5′6″	5′7″	5′8″	5′9″	5′10″
M	1.47	1.5	1.52	1.55	1.57	1.6	1.62	1.65	1.67	1.7	1.72	1.75	1.77
Age													
15y	379	382	385	389	391	394	397	400	402	405	407	411	413
20y	402	406	409	413	416	419	422	425	428	431	434	437	439
25y	415	419	422	426	429	433	435	439	441	445	447	451	453
30y	419	424	427	431	433	437	440	444	446	450	452	456	458
35y	418	423	425	430	432	436	439	443	445	449	451	454	457
40y	413	417	420	424	427	431	433	437	439	443	445	449	451
45y	405	409	412	416	418	422	425	428	431	434	436	440	442
50y	394	399	401	405	407	411	414	417	419	423	425	428	430
55y	383	387	389	393	395	399	401	404	407	410	412	415	417
60y	370	373	376	379	382	385	387	391	393	396	398	401	403
65y	356	360	362	366	368	371	373	376	378	381	383	386	388
70y	343	346	348	351	353	356	358	361	363	366	368	371	372

Men

Height: →

Feet	5′2″	5′3″	5′4″	5′5″	5′6″	5′7″	5′8″	5′9″	5′10″	5′11″	6′	6′1″	6′2″
M	1.57	1.6	1.62	1.65	1.67	1.7	1.72	1.75	1.77	1.8	1.82	1.85	1.87
Age													
15y	479	485	489	494	498	503	506	511	515	520	523	528	531
20y	534	540	545	551	555	561	565	571	575	580	584	589	593
25y	568	575	580	587	591	598	602	608	612	618	622	628	632
30y	587	594	599	606	611	617	622	628	633	639	643	649	653
35y	594	601	606	613	618	625	629	636	640	646	650	657	661
40y	592	599	604	611	615	622	627	633	637	644	648	654	658
45y	582	590	594	601	606	612	617	623	627	634	638	644	647
50y	568	575	580	586	591	597	601	608	612	618	622	627	631
55y	550	557	561	568	572	578	582	588	592	598	602	607	611
60y	529	536	540	546	550	556	560	566	570	575	579	584	588
65y	507	513	517	523	527	533	536	542	545	551	554	559	562
70y	484	490	493	499	503	508	511	517	520	525	528	533	536

❗ For normal values in age groups/heights not represented on these charts or for conversion from the old Wright scale peak flow meters see 🖱 www.peakflow.com

Source: data from Gregg I., Nunn AJ, *BMJ* 1989;**298**:1068–70 and Godfrey S, et al. *Br J Dis Chest* 1970;**64**:15.

Table 10.5 Predicted FEV_1 and FVC measurements (in L)

❗ These values apply for Caucasians. ↓ values by 7% for Asians and 13% for people of Afro-Caribbean origin

Height	Feet	4′11″	5′1″	5′3″	5′5″	5′7″	5′9″	5′11″
	Metres	1.5	1.55	1.6	1.65	1.7	1.75	1.8
Age (y) Women								
38–41	FEV_1	2.3	2.5	2.7	2.89	3.09	3.29	3.49
	FVC	2.69	2.91	3.13	3.35	3.58	3.80	4.02
42–45	FEV_1	2.2	2.4	2.6	2.79	2.99	3.19	3.39
	FVC	2.59	2.81	3.03	3.25	3.47	3.69	3.91
46–49	FEV_1	2.1	2.3	2.5	2.69	2.89	3.09	3.29
	FVC	2.48	2.7	2.92	3.15	3.37	3.59	3.81
50–53	FEV_1	2	2.2	2.4	2.59	2.79	2.99	3.19
	FVC	2.38	2.6	2.82	3.04	3.26	3.48	3.71
54–57	FEV_1	1.9	2.1	2.3	2.49	2.69	2.89	3.09
	FVC	2.27	2.49	2.72	2.94	3.16	3.38	3.6
58–61	FEV_1	1.8	2	2.2	2.39	2.59	2.79	2.99
	FVC	2.17	2.39	2.61	2.83	3.06	3.28	3.5
62–65	FEV_1	1.7	1.9	2.1	2.29	2.49	2.69	2.89
	FVC	2.07	2.29	2.51	2.73	2.95	3.17	3.39
66–69	FEV_1	1.6	1.8	2	2.19	2.39	2.59	2.79
	FVC	1.96	2.18	2.4	2.63	2.85	3.07	3.29

For women ≤70y use the formulae:

- $FEV_1 = (0.0395 \times \text{height in m} \times 100) - (0.025 \times \text{age in y}) - 2.6$
- $FVC = (0.0443 \times \text{height in m} \times 100) - (0.026 \times \text{age in y}) - 2.89$

Height	Feet	5′3″	5′5″	5′7″	5′9″	5′11″	6′1″	6′3″
	Metres	1.6	1.65	1.7	1.75	1.8	1.85	1.9
Age (y) Men								
38–41	FEV_1	3.2	3.42	3.63	3.85	4.06	4.28	4.49
	FVC	3.81	4.1	4.39	4.67	4.96	5.25	5.54
42–45	FEV_1	3.09	3.3	3.52	3.73	3.95	4.16	4.38
	FVC	3.71	3.99	4.28	4.57	4.86	5.15	5.43
46–49	FEV_1	2.97	3.18	3.4	3.61	3.83	4.04	4.26
	FVC	3.6	3.89	4.18	4.47	4.75	5.04	5.33
50–53	FEV_1	2.85	3.07	3.28	3.5	3.71	3.93	4.14
	FVC	3.5	3.79	4.07	4.36	4.65	4.94	5.23
54–57	FEV_1	2.74	2.95	3.17	3.38	3.6	3.81	4.03
	FVC	3.39	3.68	3.97	4.26	4.55	4.83	5.12
58–61	FEV_1	2.62	2.84	3.05	3.27	3.48	3.7	3.91
	FVC	3.29	3.58	3.87	4.15	4.44	4.73	5.02
62–65	FEV_1	2.51	2.72	2.94	3.15	3.37	3.58	3.8
	FVC	3.19	3.47	3.76	4.05	4.34	4.63	4.91
66–69	FEV_1	2.39	2.6	2.82	3.03	3.25	3.46	3.68
	FVC	3.08	3.37	3.66	3.95	4.23	4.52	4.81

For men ≤70y use the formulae:

- $FEV_1 = (0.043 \times \text{height in m} \times 100) - (0.029 \times \text{age in y}) - 2.49$
- $FVC = (0.0576 \times \text{height in m} \times 100) - (0.026 \times \text{age in y}) - 4.34$

Source: data from the British Thoracic Society.

Bronchodilators and steroids

Bronchodilators Cause relaxation of bronchial smooth muscle.

Short-acting β2 ***agonists*** e.g. salbutamol, terbutaline. Safest, most effective β_2 agonists for use as quick relievers in asthma and COPD.

- Duration of action: ~3–5h. Oral preparations are less effective than inhaled preparations. Prescribe as 1–2 puffs prn
- Warn patients to seek medical advice if usual dose does not relieve symptoms or relieves symptoms for <3h
- Regular treatment with bronchodilators alone may be linked with worsening of asthma and asthma deaths. If the patient has asthma and is using a β_2 agonist inhaler >3×/wk, consider prophylaxis—➔ p. 282

Longer-acting β2 agonists e.g. salmeterol, formoterol.

- Asthma e.g. salmeterol 50–100mcg bd as an adjunct to existing corticosteroid treatment—➔ p. 282. Particularly useful for night-time asthma. Duration of action is ~12h. Due to its rapid onset of action, formoterol can also be used for relief of acute attacks, usually in combined steroid/LABA inhaler, as part of a maintenance and reliever therapy (MART) regime.
- COPD ➔ p. 286

❗ Always prescribe as combination inhaler with inhaled steroid.

Steroids Short- and long-term treatment of inflammatory conditions.

- Oral steroids Prescribe as a single dose in the morning. Often started at high dose (e.g.40–50mg od) to suppress disease process and then stopped after improvement. If used as maintenance therapy, use the minimum dose that controls disease. Supply with a 'steroid card'
- Inhaled steroids Use regularly to obtain maximum benefit. Alleviation of symptoms occurs 3–7d after initiation. If causes coughing, try a short-acting β_2 agonist before use. Common unwanted effects are oral candidiasis (5%) and hoarseness—↓ by use of a large volume spacer or mouth washing after use

❗ Beclometasone inhalers should always be prescribed by brand name.

Side effects of oral and high-dose inhaled steroids

- ↑ BP
- Osteoporosis ± fracture
- Proximal muscle wasting
- Euphoria
- Paranoid states/depression—especially if PMH
- Peptic ulceration—po soluble or EC preparations may ↓ risk
- Suppression of clinical signs—may allow diseases, e.g. septicaemia, to reach advanced stage before being recognized
- Spread of infection, e.g. chickenpox
- DM/worsening of diabetic control
- Cushing's syndrome—moon face, striae, and acne
- Adrenal atrophy—can persist years after stopping long-term steroids—illness/surgical emergencies may need to be covered with steroid supplements
- Growth suppression in children
- Na^+ and water retention; K^+ loss

Steroid cards Should be carried by patients on oral/high doses of inhaled steroids. The card informs other practitioners that the patient is on steroids and gives the patient advice on use of steroids and risk of infection. Steroid cards can be obtained from: ☎ 0161 6832189 Email: nhsforms@spsl.uk.com

Withdrawal of steroids Stop abruptly if disease is unlikely to relapse, the patient has received treatment for ≤3wk and is not included in the following patient groups. Withdraw gradually if disease is unlikely to relapse and the patient has:

- Recently had repeated steroid courses (particularly if taken for >3wk)
- Taken a short course <1y after stopping long-term therapy
- Other possible causes of adrenal suppression
- Received >40mg od of prednisolone (or equivalent) for >1wk
- Been given repeat doses in the evening
- Received treatment with steroids for >3wk

During corticosteroid withdrawal, ↓ dose rapidly to physiological levels (~prednisolone 7.5mg od)—thereafter ↓ more slowly. Assess the disease during withdrawal to ensure relapse does not occur.

Use of spacers with metered dose inhalers (MDIs)

Advantages of using a spacer Allows more time for evaporation of propellant so a larger proportion of active drug is deposited in the lungs; there is no need to coordinate actuation with inhalation; results in less oropharyngeal side effects (e.g. thrush, hoarseness with inhaled steroids).

Use of spacers Both large-volume spacers (e.g. Volumatic™) and medium-volume devices (e.g. Aerochamber™) are widely available, acceptable, and portable. Inhale the drug from the spacer immediately after actuation as effect of the drugs is short-lived. Spacers should be washed and air dried monthly to prevent build-up of electrostatic charge affecting drug delivery, and replaced every 6–12mo.

Home nebulizer therapy In England and Wales nebulizers are not available via the NHS (but are free of VAT). Some nebulizers are available in Scotland on form GP10A. Nebulizers convert a solution of drug into an aerosol for inhalation. They are used to deliver a higher dosage of drug than is usual with inhalers over a short period of time (5–10min). *Indications*:

- Acute exacerbations ± regular treatment of asthma/COPD
- Antibiotic treatment—for patients with chronic purulent infection, e.g. CF; bronchiectasis; prophylaxis and treatment of pneumocystis pneumonia with pentamidine in patients with AIDS
- Palliative care—palliation of breathlessness and cough, e.g. bronchodilators, lidocaine, or bupivacaine for dry, persistent cough

Use in asthma/COPD Before suggesting long-term use:

- Review diagnosis, technique using hand-held device ± spacer, and compliance
- Try ↑ dose of bronchodilator via a hand-held device for at least 2wk
- Perform a 2wk trial of nebulizer therapy and monitor therapeutic effect (e.g. with PEFR in asthma, or dyspnoea score with COPD)
- Provide clear instructions on the use of the nebulizer, monitoring, and when to seek help. Follow up regularly

Further information

European Respiratory Society (2001) Guidelines on the use of nebulizers. *Eur Respir J* **18**:228–42.

Asthma in adults

Symptoms/signs of a severe asthma attack

- PEFR 33–50% predicted or best
- Oxygen saturation ≥92%
- Unable to talk in sentences
- Tachypnoea (respiratory rate ≥25 breaths/min)
- Tachycardia (heart rate ≥110bpm)

Life-threatening signs

- PEFR <33% predicted or best
- Oxygen saturation <92%
- Poor respiratory effort
- Silent chest (inaudible wheeze)
- Arrhythmia
- Hypotension
- Altered consciousness
- Cyanosis
- Exhaustion

Management of an acute asthma attack ➔ p. 1072

💣 There are two competing national asthma guidelines in the UK which differ significantly from each other. The guidance in this section is based on BTS/SIGN guidance; links to both the BTS/SIGN guidance and parallel NICE guidance are provided on ➔ p. 281.

Asthma is a condition of paroxysmal, reversible airways obstruction that affects 1 in 12 adults in the UK. It has 3 characteristic features:

- Airflow limitation—usually reversible spontaneously or with treatment
- Airway hyper-responsiveness to a wide range of stimuli
- Inflammation of the bronchi

Asthma in special groups

- Children ➔ p. 860
- Occupational asthma ➔ p. 306
- Pregnancy ➔ p. 800

Diagnosis of asthma Characteristic pattern of symptoms/signs and tests without another explanation. Determine probability of asthma:

- High Recurrent, episodic asthma symptoms (>1 of wheeze, breathlessness, chest tightness, cough), diurnal variability, audible expiratory wheeze, documented variable airflow obstruction (e.g. PEFR variability), atopic history, absence of features to suggest another diagnosis
- Intermediate Some features of asthma; poor treatment response
- Low No typical asthma features; symptoms suggest another diagnosis

❗ Always consider the possibility of occupational asthma[N]. Ask: '*Are symptoms better on days away from work?*' and '*Are symptoms better on holiday?*' If yes to either/both, consider referral.

Clinical features that ↓ probability of asthma

- Normal chest examination and/or PEFR/spirometry when symptomatic
- Chronic productive cough without wheeze/breathlessness
- Prominent dizziness, light-headedness, peripheral tingling
- Voice disturbance
- Symptoms with colds only
- Smoking history (>20 pack y)
- Cardiac disease

❗ Normal spirometry when asymptomatic does not exclude asthma.

Tests Spirometry + reversibility is the preferred initial test; consider CXR with atypical/additional symptoms; eosinophil counts. ❗ NICE advocates initial use of fractional exhaled nitric oxide (FeNO) testing.

Differential diagnosis Airflow obstruction = FEV$_1$/FVC <0.7

Airflow obstruction

- COPD
- Bronchiectasis*
- Inhaled foreign body*
- Obliterative bronchiolitis
- Large airway stenosis
- Lung cancer*
- Sarcoidosis*

No airflow obstruction

- Chronic cough syndromes
- Hyperventilation syndrome
- Vocal cord dysfunction
- Rhinitis
- Gastro-oesophageal reflux
- Heart failure
- Pulmonary fibrosis

*May also be associated with non-obstructive spirometry.

Action

High probability Give trial of treatment with inhaled beclometasone 200mcg bd (or equivalent) for 6wk. If response is poor despite adequate inhaler technique/concordance, investigate further.

❗ If significant airflow obstruction, there may be inhaled steroid resistance. Treat with oral prednisolone 30mg od for 2wk instead.

Intermediate probability Perform spirometry:

- If FEV$_1$/FVC <0.7 (i.e. airways obstruction present)—offer reversibility testing and/or trial of treatment. If significant reversibility and/or trial of treatment is beneficial, treat as asthma. If insignificant reversibility and treatment trial is not beneficial, consider tests for alternative diagnoses
- If FEV$_1$/FVC >0.7 (i.e. no evidence of airways obstruction), arrange further investigations, e.g. challenge tests or FeNO testing to identify eosinophilic inflammation (≥40 parts per billion is +ve indicating high likelihood of asthma), before commencing treatment ± refer

Low probability Consider alternative diagnoses and investigate/manage accordingly. Reconsider asthma if no response.

Reversibility testing For patients with diagnostic uncertainty:

- If airflow obstruction is present at the time of assessment—assess FEV$_1$ (or PEFR) and/or symptoms before and 20min after 400 mcg inhaled salbutamol via MDI and spacer
- If no airflow obstruction is present, or response to inhaled salbutamol is uncertain—assess FEV$_1$ (or PEFR) and/or symptoms after trial of treatment with inhaled beclometasone 200mcg bd or equivalent for 6wk, or oral steroids (prednisolone 30mg od for 14d)

>200mL ↑ in FEV$_1$ suggests (>400mL ↑ strongly suggests) asthma. If smaller improvement, decide whether to continue treatment by assessment of symptoms. Trial of treatment withdrawal may be helpful if doubt.

Reasons for referral *E* = Emergency; *U* = Urgent; *S* = Soon; *R* = Routine

- Severe asthma exacerbation *E*
- Monophonic wheeze/stridor *E/U*
- CXR shadowing *U*
- Prominent systemic features (myalgia, fever, weight loss) *U/S*
- Diagnosis unclear *S/R*
- Unexpected clinical findings (e.g. crackles, clubbing, cyanosis) *S/R*
- Constant breathlessness *S/R*
- Poor response to treatment *S/R*
- Unexplained restrictive spirometry *R*
- Suspected occupational asthma *R*
- Chronic sputum production *R*
- Eosinophilia ($>1 \times 10^9$/L) *R*

Asthma management in practice

💣 There are two competing national asthma guidelines in the UK which differ significantly from each other. The guidance in this section is based on BTS/SIGN guidance; a link to the parallel NICE guidance is provided.

Aims of treatment To:
- ↓ daytime symptoms and night-time waking due to asthma
- Minimize the need for reliever medication
- ↓ impact on lifestyle, e.g. absences from work/school, exercise
- Prevent severe attacks/exacerbations
- Have normal lung function (FEV_1 and/or PEF >80% predicted or best)
- ↓ side effects from medications

GP services Routine asthma care should be carried out in a specialized primary care clinic. Doctors/nurses involved need appropriate training and regular updates. Practices should keep an asthma register to ensure adequate follow-up and allow audit.

❗ Not all patients want to attend a pre-arranged appointment. Telephone reviews may be as effective as face-to-face consultations.

Reviews and monitoring Frequency depends on needs. Aim to review all patients with asthma at least annually (Figure 10.2).
- Check symptoms since last seen. Use objective measures, e.g. RCP 3 questions (➔ p. 273)
- Record smoking status and advise smokers to stop

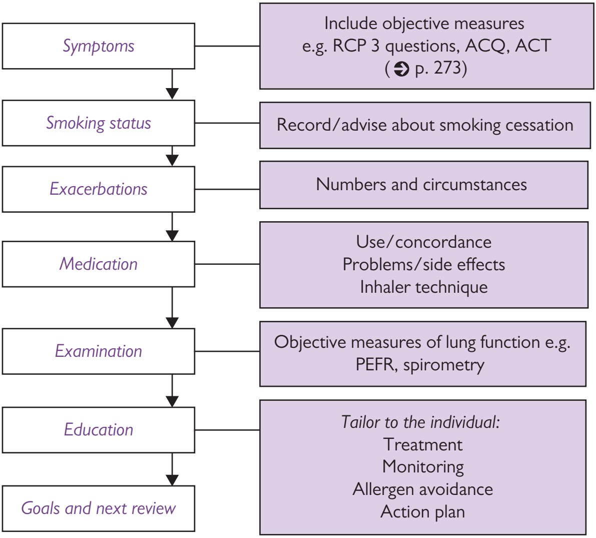

Symptoms
Include objective measures e.g. RCP 3 questions, ACQ, ACT (➔ p. 273)
Smoking status
Record/advise about smoking cessation
Exacerbations
Numbers and circumstances
Medication
Use/concordance
Problems/side effects
Inhaler technique
Examination
Objective measures of lung function e.g. PEFR, spirometry
Education
Tailor to the individual:
Treatment
Monitoring
Allergen avoidance
Action plan
Goals and next review

Figure 10.2 Summary of the annual asthma review

- Record any exacerbations/acute attacks since last seen
- Check medication—use, concordance (prescription count—➔ p. 116), inhaler technique, problems, side effects. ❗ If >1 SABA inhaler/mo, may have poor asthma control—consider stepping up treatment
- Check influenza/pneumococcal vaccination received
- Review objective measures of lung function, e.g. home PEFR chart, PEFR/spirometry at review
- Address any problems or queries and educate about asthma
- Agree management goals and date for further review

Self-management All patients should receive:

- Self-management education Brief, simple education linked to patient goals is most likely to be successful. Include information about: nature of disease, nature of the treatment and how to use it, self-monitoring/self-assessment, recognition of acute exacerbations, allergen/trigger avoidance, patients' own goals of treatment
- Written action plan Focus on individual needs. Include information about symptom triggers and peak flow levels that indicate when asthma is worsening, and guidance about what to do under those circumstances. Action plans ↓ morbidity and health costs from asthma
- PEFR monitoring Record PEFR at asthma review and if acute exacerbation. Home monitoring + action plan can be useful especially for patients with severe asthma, brittle asthma (i.e. rapid development of acute asthma attacks), and/or if poor perceivers of symptoms

Management of acute asthma ➔p. 1072

Non-pharmacological measures

- Smoking May ↑ symptoms of asthma—advise to stop
- Weight There is some evidence that weight ↓ in obese patients with asthma results in ↑ asthma control
- Allergen avoidance
  - *House dust mite*: there is little evidence that ↓ house dust mite results in clinical improvement. Physical and chemical methods of reducing house dust mite levels are ineffective and should not be recommended, e.g. acaricides, mattress covers, vacuum cleaning, heating, ventilation, freezing, washing, air filtration, and ionizers
  - *Pets*: there is no evidence that removing pets from a home results in improved symptoms, but many experts still advise removal of the pet if patients with asthma also have an allergy to the pet

Drug therapy ➔ p. 282

Further information

BTS/SIGN (2019) British guideline to the management of asthma. 🖱 www.sign.ac.uk/sign-158-british-guideline-on-the-management-of-asthma.html
NICE (2017) Asthma: diagnosis, monitoring and chronic asthma management. 🖱 www.nice.org.uk/guidance/ng80

Patient information and support

Asthma UK ☎ 0300 222 5800 🖱 www.asthma.org.uk

Drug treatment of asthma

💣 There are two competing national asthma guidelines in the UK which differ significantly from each other. The guidance in this section is based on BTS/SIGN guidance; a link to the parallel NICE guidance is provided.

Management of acute asthma ➲ p. 1072

Exacerbations Treat early. In adult patients on 200mcg doses of inhaled steroids, a 5× ↑ in dose reduces severity of exacerbations. Alternatively, use prednisolone 30–40mg od for 1–2wk.

Use a stepwise approach Figure 10.3. Start at the step most appropriate to the initial severity of symptoms. Achieve early control. Maintain control by ↑ treatment if needed and ↓ treatment if good control.

Selection of inhaler device If possible, use a MDI. Inadequate technique may result in drug failure. Patients must inhale slowly and hold their breath for 10sec after inhalation. Demonstrate inhaler technique before prescribing and check at follow-ups. Spacers/breath-activated devices are useful if patients find activation difficult. Dry powder inhalers are an alternative.

Short-acting β_2 agonists ➲ p. 276—e.g. salbutamol/terbutaline. Work more quickly and with fewer side effects than alternatives. Use prn unless shown to benefit from regular dosing. Using ≥2 canisters/mo or >10–12 puffs/d is a marker of poorly controlled asthma.

❗ A budesonide/formoterol combination inhaler is an alternative rescue medication as part of a MART regime.

Inhaled corticosteroids ➲ p. 276—effective preventer. May be beneficial even for patients with mild asthma. Consider if: exacerbation of asthma in the last 2y requiring steroids; using inhaled β_2 agonists ≥3×/wk; or symptomatic ≥3×/wk or ≥1 night/wk.

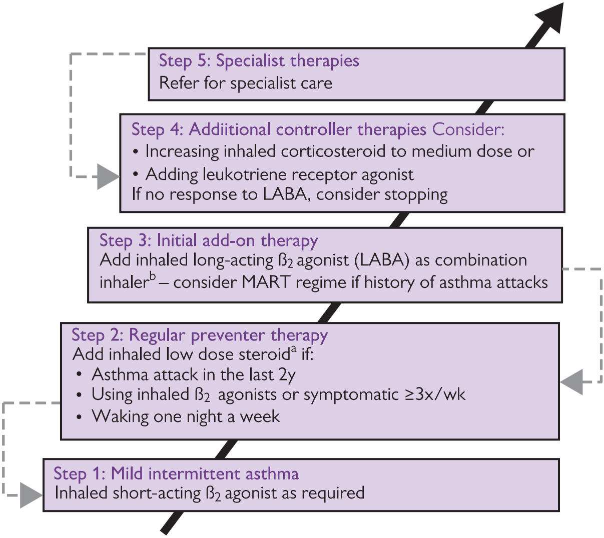
Oral steroids ➲ p. 276

Add on therapy Aims to improve lung function/symptoms. Before initiating a new drug, check compliance, inhaler technique, and eliminate trigger factors. Only continue if of demonstrable benefit.

- Inhaled long-acting β_2 agonists (LABA) ➲ p. 276—e.g. salmeterol. Although there is no difference in efficacy, combination inhalers are recommended to improve inhaler concordance. A MART regime using a steroid/formoterol combined inhaler for both regular prevention and relief of acute symptoms is useful to ↓ asthma attacks.
- Leukotriene receptor agonists e.g. montelukast. ↓ exacerbations
- Theophylline Side effects are common e.g. nausea, gastric irritation

Complementary therapies Buteyko breathing technique ↓ symptoms. No convincing evidence of effectiveness of any other therapies.

Monocloncal antibodies e.g. omalizumab, mepolizumab. May be considered in patients with a high oral corticosteroid burden. Always specialist initiated. Side effects include local skin reactions and anaphylaxis.



Step 5: Specialist therapies
Refer for specialist care

Step 4: Addiitional controller therapies Consider:
- Increasing inhaled corticosteroid to medium dose or
- Adding leukotriene receptor agonist

If no response to LABA, consider stopping

Step 3: Initial add-on therapy
Add inhaled long-acting β_2 agonist (LABA) as combination inhaler[b] – consider MART regime if history of asthma attacks

Step 2: Regular preventer therapy
Add inhaled low dose steroid[a] if:
- Asthma attack in the last 2y
- Using inhaled β_2 agonists or symptomatic ≥3x/wk
- Waking one night a week

Step 1: Mild intermittent asthma
Inhaled short-acting β_2 agonist as required

Start treatment at the level most appropriate to initial severity.
Achieve early control.
Maintain control by increasing treatment as necessary, and decreasing treatment when control is good.
Before initiating any new drug therapy, check adherence and inhaler technique and eliminate trigger factors

[a] Low dose is defined as the lowest dose recommended for an adult for the drug chosen.
[b] NICE recommends adding a leukotriene receptor agonist before LABA as initial add-on therapy.

Figure 10.3 Summary of stepwise management in adults

Source: data from British Thoracic Society and Scottish Intercollegiate Guidelines Network, SIGN 158: British guideline on the management of asthma (revised 2019) https://www.sign.ac.uk/assets/sign158.pdf

Stepping down Review and consider stepping down at intervals ≤3 mo. Maintain on the lowest dose of inhaled steroid controlling symptoms. When reducing steroids, cut dose by 25–50% each time.

Difficult asthma Persistent symptoms and/or frequent exacerbations despite treatment at step 4/5. Check diagnosis and exacerbating factors. Assess adherence to medication. Find out about family, psychological, or social problems that may be interfering with effective management.

Further information

BTS/SIGN (2019) British guideline to the management of asthma. ✆ www.sign.ac.uk/sign-158-british-guideline-on-the-management-of-asthma.html
NICE (2017) Asthma: diagnosis, monitoring and chronic asthma management. ✆ www.nice.org.uk/guidance/ng80

Chronic obstructive pulmonary disease

Chronic obstructive pulmonary disease (COPD) is a slowly progressive disorder characterized by airflow obstruction. In the UK, it affects ~3million people, with two-thirds undiagnosed. ♂ > ♀. Responsible for 1 in 8 emergency hospital admissions and ~5% of deaths in the UK[N]. *Causes*:

- Tobacco smoking (90% of cases)
- Occupational exposure, e.g. coal, silica, welding fumes
- Air pollution
- Genetic—bronchial hyper-responsiveness; α1-antitrypsin deficiency
- Poor lung growth and development e.g. low birth weight, infections

Incidental CXR/CT scan abnormalities ➔ p. 272

Diagnosis Suggested by history, signs, and baseline spirometry. Consider in patients >35y with a risk factor for COPD (generally smoking) and typical symptoms:

- Shortness of breath on exertion—use an objective measure, e.g. MRC dyspnoea scale (Table 10.6) to grade breathlessness
- Chronic cough
- Wheeze
- Regular sputum production
- Frequent winter 'bronchitis'

Table 10.6 MRC Dyspnoea Scale

Grade	Degree of breathlessness related to physical activity
1	Not troubled by breathlessness except on strenuous exercise
2	Short of breath when hurrying or walking up a slight hill
3	Walks slower than contemporaries on level ground because of breathlessness or has to stop for breath when walking at own pace
4	Stops for breath after walking 100m or after a few minutes on level ground
5	Too breathless to leave the house or breathless on dressing/undressing.

Used with the permission of the Medical Research Council.

If diagnosis is suspected also ask about Weight ↓, effort intolerance, waking at night, ankle swelling, fatigue, and occupational hazards.

⚠ Chest pain or haemoptysis are uncommon in COPD—if present consider an alternative diagnosis.

Signs May be none. *Possible signs*:

- Hyperinflated chest ± poor chest expansion on inspiration
- ↓ crico-sternal distance
- Hyper-resonant chest with ↓ cardiac dullness on percussion
- Wheeze or quiet breath sounds
- Paradoxical movement of lower ribs
- Use of accessory muscles
- Tachypnoea
- Pursing of lips on expiration (pursed lip breathing)
- Peripheral oedema
- Cyanosis
- ↑ JVP
- Cachexia

Spirometry ➔ p. 272. Predicts severity and prognosis but not disability/quality of life. Measured post-bronchodilator, e.g. 200mcg salbutamol via spacer. Diagnose airflow obstruction if FEV$_1$/FVC <0.7 (<70%). Grade severity according to the reduction in FEV$_1$ (Table 10.7).

Table 10.7 Severity of COPD and expected clinical picture

Severity	Spirometry (FEV1/FVC <0.7)	Clinical picture
Stage 1—mild	FEV$_1$ ≥80%	**Cough.** Little/no breathlessness. No abnormal signs. No ↑ use of services
Stage 2—moderate	FEV$_1$ 50–79% predicted	Breathlessness, wheeze on exertion, cough ± sputum. Some abnormal signs. Known to GP—intermittent problems
Stage 3—severe	FEV$_1$ 30–49% predicted	**SOBOE.** Marked wheeze/cough. Usually other signs. Known to GP and specialist with frequent problems/admissions
Stage 4—very severe	FEV$_1$ <30% predicted	As for stage 3 but more breathless; severely restricted activities of daily living

Reversibility testing Can be misleading. Not routinely recommended:

- >400mL ↑ in FEV$_1$ following trial of bronchodilator or prednisolone (30mg od for 2wk) suggests asthma
- Clinically significant COPD is *not* present if FEV$_1$ and FEV$_1$/FVC return to normal after drug therapy

PEFR ➔ p. 272. Patients with COPD have little variability in PEFR. Serial home PEFR measurements can help distinguish between asthma and COPD. PEFR may underestimate severity of airflow limitation and a normal PEFR does not exclude airflow obstruction.

Other investigations organized in primary care

- **CXR/CT** Indicated to exclude other diagnoses e.g. lung cancer
- **FBC** To identify secondary polycythaemia or anaemia
- **BMI**
- **α1-antitrypsin** If early onset COPD or family history—➔ p. 398
- **ECG/Echo/serum natriuretic peptide** If cor pulmonale is suspected
- **Sputum culture** If purulent sputum is persistent
- **Pulse oximetry** To assess need for oxygen therapy

Differential diagnosis Asthma (Table 10.8), bronchiectasis, heart failure, lung cancer, interstitial lung disease, anaemia, tuberculosis. Refer for specialist review if diagnostic uncertainty.

Table 10.8 Comparison of COPD and asthma

	COPD	Asthma
Symptoms <35y	Rare	Often
Smoking history	Nearly all	Maybe
Breathlessness	Persistent and progressive. Poor response to inhaled therapy—if good reconsider diagnosis	Variable throughout the day and from day to day. Good response to inhaled therapy is typical
Chronic productive cough	Common	Uncommon
Waking at night with cough/wheeze	Uncommon	Common

Management of COPD

Develop an individualized management plan for each patient. Review ≥1×/y. Record spirometry results and progression, BMI, current symptoms, problems since last seen, exercise tolerance, and smoking status. Educate patient/family about COPD, medication, and self-help.

Non-drug therapy

- **Smoking cessation** Most important. Improves outcome (Table 10.9)
- **Vaccination** All patients with COPD should have influenza and pneumococcal vaccination
- **Pulmonary rehabilitation** Lack of exercise ↓ FEV_1. Multidisciplinary pulmonary rehabilitation is of proven benefit—refer for patients who consider their COPD hinders everyday activities
- **Nutrition** Weight ↓ in obese patients improves exercise tolerance
- **Cognitive behavioural techniques** Can help patients who find the sensation of being breathless frightening
- **Screening for depression** ➔ p. 173

Table 10.9 Smoking and COPD

	FEV_1 as % of value aged 25y	
	Age 60y	Age 75y
Non-smoker	85%	80%
Ex-smoker: quit aged 40y	60%	45% (symptoms)
Ex-smoker: quit aged 60y	33% (severe symptoms)	15% (severe disability)
Ongoing smoker	33% (severe symptoms)	Dead

Rescue medication and escalation plans ➔ p. 288

Management of acute exacerbations ➔ p. 288

Long-term drug therapy Document effects of each drug treatment on symptoms, quality of life, and lung function as tried.

- **Step 1** Offer a short-acting β2 agonist (SABA) e.g. salbutamol inhaler 1–2 puffs as needed, or short-acting muscarinic antagonist (SAMA) e.g. ipratropium 20–40mcg. Both ↑ FEV_1 and ↓ breathlessness
- **Step 2** Add:
  - *If asthma features* (i.e. some steroid responsiveness). Long-acting β2 agonist (LABA) + inhaled corticosteroid (e.g. fluticasone/vilanterol)
  - *If no asthma features* Long-acting muscarinic antagonist (LAMA)+ LABA (e.g. vilanterol/umeclidinium)
- **Step 3** If asthma features/steroid responsiveness, continue short-acting bronchodilator and change long-acting medication to a LABA + LAMA + inhaled corticosteroid inhaler (e.g. fluticasone/vilanterol/umeclidinium)

Spacer devices ➔ p. 277 **Nebulizers** ➔ p. 277

Add-on therapy

Oral mucolytic therapy e.g. carbocisteine. Consider if chronic cough + sputum. Discontinue if no symptomatic improvement.

Oral prophylactic antibiotics^{N} Azithromycin 250mg 3×/wk. Offer if non-smoker/ex-smoker, optimized inhaled therapies, and has undertaken pulmonary rehabilitation but frequent (≥4/y)/prolonged exacerbations with sputum production, or hospitalizations. Before starting, check sputum for M,C&S to exclude resistant organisms/*Pseudomonas aeruginosa*, baseline LFTs, and ECG (for prolonged QT interval). Advise small risk of hearing loss/tinnitus. Review after 3mo and then every 4–6mo. Only continue if benefits > risks. Doxycycline 100mg od is an alternative.

Long-term oral steroids Avoid if possible. If unavoidable keep dose as low as possible and provide bone protection (➜ p. 484).

Theophylline M/R Only use if inhaled LABA/LAMA is ineffective or unable to use inhaled medication. Monitoring of serum drug levels is required. Be wary of drug interactions—especially with antibiotics.

Roflumilast Specialist initiation only. Adjunct to bronchodilators for patients with severe COPD with a history of frequent exacerbations.

Referral for specialist care

- Uncertain diagnosis
- Age <40y
- Severe COPD
- Rapid decline in FEV$_1$
- Haemoptysis—urgent referral
- Frequent exacerbations
- Pulmonary hypertension/cor pulmonale
- α1-antitrypsin deficiency
- Assessment for: LTOT, withdrawal of long term steroids, long-term nebulizer therapy, surgery, e.g. lung transplant, bullectomy

Long-term oxygen therapy (LTOT) *Only* prescribe after evaluation by a respiratory physician. Refer patients with:

- Severe airflow obstruction (FEV$_1$ <30%—consider if 30–49%)
- Hypoxaemia (oxygen saturation ≤92% breathing air)
- Cyanosis
- Polycythaemia
- Peripheral oedema
- ↑ JVP

Treatment for >15h/d ↑ survival and quality of life. Ambulatory oxygen can ↑ exercise tolerance in some patients. ❗ Always warn patients about the fire risks of having pure oxygen in their homes.

O_2 cylinders and associated equipment Arrangements for supply of oxygen are differ across the UK—see BNF. Specify amount of O_2 required (h/d) and flow rate. O_2 concentrators are more economical for LTOT. Supply back up cylinders in case of breakdown or power cut.

Prognosis Poor prognosis is associated with:

- Smoking status (smoker)
- Low BMI
- Chronic hypoxia and/or cor pulmonale
- Multimorbidity/frailty
- Severe/frequent exacerbations ± hospital admissions
- Breathless (MRC4/5), high symptom burden ± ↓ exercise capacity
- Low FEV$_1$ and/or meets criteria for LTOT

Cor pulmonale ➜ p. 236

Palliative care ➜ p. 1011

Further information

NICE (2018) Chronic obstructive pulmonary disease in over 16s: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng115

Patient support

British Lung Foundation ☎ 03000 030 555 🖱 www.lunguk.org

Acute exacerbations of COPD

Risk factors for exacerbation

- Exposure to passive smoke
- Viral or bacterial infection
- Lack of physical activity
- Seasonal variation (winter and spring)
- Continued smoking or relapse for ex-smokers
- Indoor and outdoor air pollution, e.g. nitrous oxide, ozone

Presentation Worsening of previous stable condition. *Features*: ≥1 of

- ↑ dyspnoea—marked dyspnoea, tachypnoea (>25 breaths/min), use of accessory muscles at rest and pursed lip breathing are signs of severe exacerbation
- ↓ exercise tolerance—marked ↓ in activities of daily living is a sign of severe exacerbation
- ↑ fatigue
- ↑ fluid retention—new onset oedema is a sign of severe exacerbation
- ↑ wheeze
- Chest tightness
- ↑ cough
- ↑ sputum purulence
- ↑ sputum volume
- Upper airways symptoms, e.g. cold, sore throat
- New-onset cyanosis—severe exacerbation
- Acute confusion—severe exacerbation

❗ Fever and chest pain are uncommon presenting features—consider alternative diagnosis.

Differential diagnosis

- Pneumonia
- LVF/pulmonary oedema
- Lung cancer
- Pleural effusion
- Recurrent aspiration
- Pneumothorax
- PE
- Upper airway obstruction

Investigations

- **Pulse oximetry** Can be used to assess severity (saturation ≤92% on air suggests hypoxaemia—consider admission) and to monitor progress
- **CXR** Consider if diagnostic doubt and/or to exclude other causes of symptoms
- **Sputum culture** Not recommended routinely in the community

Rescue medication and escalation plans Develop an individualized escalation plan with each patient to manage acute exacerbations, If able to understand their use, offer patients at risk of exacerbations a short course of oral steroids and antibiotics to keep at home to enable prompt treatment of exacerbations.

❗ Review if using ≥3 courses of oral steroids and/or antibiotics per year.

GP management Decide whether to treat at home or admit to hospital—Table 10.10. If managing in the community:

- **Add or ↑ bronchodilators** Consider if inhaler device and technique are appropriate
- **Oral corticosteroids** Start early in the course of the exacerbation if ↑ breathlessness which interferes with daily activities. Dosage—30–40mg/d of prednisolone for 1–2wk. Consider osteoporosis prophylaxis with a bisphosphonate if frequent courses are required (➔ p. 484)

Table 10.10 Deciding to treat exacerbations at home or in hospital. The more features in the 'treat in hospital column', the more likely the need for admission

	Treat at home	Treat in hospital[a]
Ability to cope at home	Yes	No
Breathlessness	Mild	Severe
General condition	Good	Poor—deteriorating
Level of activity	Good	Poor/confined to bed
Cyanosis	No	Yes
Worsening peripheral oedema	No	Yes
Level of consciousness	Normal	Impaired
Already receiving LTOT	No	Yes
Social circumstances	Good	Living alone/not coping
Acute confusion	No	Yes
Rapid rate of onset	No	Yes
Significant co-morbidity (e.g. cardiac disease, DM)	No	Yes
SaO2 <90% on air	No	Yes
Changes on CXR (if available)	No	Present

[a] Hospital-at-home schemes and assisted discharge schemes are a suitable alternative.

Source: data from Gravil, JH, et al. Home treatment of exacerbations of chronic obstructive pulmonary disease by an acute respiratory assessment service. *Lancet* 1998; **351**(9119):1853–5.

- **Start antibiotics** Use broad-spectrum antibiotic for 5d (e.g. amoxicillin 500mg tds, erythromycin 500mg qds, clarithromycin 500mg bd, or doxycycline 200mg on day 1 then 100mg od) if sputum changes colour or ↑ in volume/thickness beyond normal day-to-day variation, clinical signs of pneumonia, or consolidation on CXR

Follow-up

- Reassess as necessary. If the patient deteriorates reconsider the need for hospital admission. If not fully improved in within 2wk consider CXR and/or hospital referral
- Reassess patients who have been admitted 4–6wk after discharge Assess their ability to cope at home. ~1 in 3 are readmitted in <3mo
- Reassess inhaler technique and understanding of treatment regimen
- In severe cases, reassess the need for LTOT and/or home nebulizer
- Check FEV_1
- Emphasize the potential benefit of lifestyle modification—smoking cessation, exercise, weight loss if obese
- Arrange ongoing regular follow-up

Further information

NICE (2018) Chronic obstructive pulmonary disease in over 16s: diagnosis and management. 🖱 https://www.nice.org.uk/guidance/ng115
NICE (2018) Chronic obstructive pulmonary disease (acute exacerbation): antimicrobial prescribing. 🖱 https://www.nice.org.uk/guidance/ng114

Lung cancer

Referral for suspected lung cancer[N]

Immediate referral/acute admission

- Stridor
- Superior vena cava obstruction (swelling of face/neck with fixed ↑ JVP)

Urgent referral To a team specializing in management of lung cancer:

- CXR suggestive of lung cancer
- ≥40y with unexplained haemoptysis

Urgent referral for CXR

- If ≥40y + ≥2 of the following unexplained symptoms (≥1 if current/previous smoker or exposure to asbestos):
  - Cough
  - Fatigue
  - Shortness of breath
  - Chest pain
  - Weight ↓
  - Appetite ↓

Consider urgent referral for CXR If

- Persistent or recurrent chest infections
- Finger clubbing
- Supraclavicular lymphadenopathy or persistent cervical lymphadenopathy
- Chest signs consistent with lung cancer
- Thrombocytosis

Lung cancer is the third most common cancer in the UK, but the most common cause of cancer death (22%). There are more than 39,000 new cases of lung cancer in the UK each year. Incidence ↑ with age—48% of lung cancer deaths each year occur in those aged ≥75y. Historically, more common in males, though 45% of new diagnoses are now in females.

Types

- **Small cell lung cancer** ~20% all cases. Often disseminated at diagnosis. Spreads to liver, bones, brain, and adrenals
- **Non-small cell lung cancer** ~80% all cases. Mainly adenocarcinoma or squamous cell carcinoma. Not always smoking-related

Screening Current evidence does not support screening for lung cancer with chest radiography or sputum cytology. Frequent chest X-ray screening may be harmful. Screening with low-dose CT was found in one trial to ↓ the number people of who died from lung cancer, but this trial included very high-risk smokers and ex-smokers. However, there were high false +ve rates. More research is needed about the use of CT screening in lower-risk individuals.

Prevention

- **Smoking cessation** ~90% of lung cancers are caused by smoking. The younger a person is when he/she starts smoking, the greater the risk of developing lung cancer. Risk also ↑ with amount smoked (duration of smoking and number of cigarettes smoked/d). Offer support and treatments to aid smoking cessation, e.g. e-cigarettes, nicotine replacement, varenicline, bupropion—➔ p. 156

- **Diet** ↑ consumption of fruit, carrots, and green vegetables may ↓ incidence but there is no evidence that vitamin supplements are beneficial and they might be harmful[C]

Presentation >90% have symptoms at the time of diagnosis. Common presenting features:

- Cough
- Chest/shoulder pain
- Haemoptysis
- Dyspnoea
- Hoarseness
- Weight ↓
- Finger clubbing
- General malaise
- Distant metastases
- Incidental finding on CXR

Pancoast syndrome Apical lung cancer + ipsilateral Horner's syndrome. *Cause*: invasion of the cervical sympathetic plexus. *Other features*: shoulder and arm pain (brachial plexus invasion C8–T2) ± hoarse voice/bovine cough (unilateral recurrent laryngeal nerve palsy and vocal cord paralysis).

Paraneoplastic syndromes e.g. ectopic ACTH production, SIADH, hypercalcaemia, hypercoagulability. Affect 10–20% of patients with lung cancer—particularly small cell. Have a high index of suspicion and refer for specialist management if suspected.

Management Once the diagnosis has been confirmed, liaise with the chest physician, specialist lung cancer team, PHCT, and specialist palliative care services (e.g. Macmillan Nurses). Active treatment options depend on type and extent of tumour and include surgery, radiotherapy, and/or chemotherapy. Follow-up regularly. 70% die in <1y. Long-term survival (>10y) is currently only ~5.5%.

Palliative radiotherapy Radiotherapy is a key component of symptomatic treatment for:

- Haemoptysis
- Chest pain
- Breathlessness due to bronchial occlusion
- Pain from bone metastasis
- Symptoms from brain metastasis

Radiotherapy may be combined with palliative chemotherapy, particularly for patients with non-small cell lung cancer.

Mesothelioma ➔ p. 307

Palliative care ➔ p. 1011

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12
NICE (2019) Lung cancer: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng122
SIGN (2014) Management of lung cancer. 🖱 www.sign.ac.uk/sign-137-management-of-lung-cancer.html

Information and support for patients

British Lung Foundation ☎ 03000 030 555 🖱 www.blf.org.uk
Cancer Research UK ☎ 0808 800 4040 🖱 www.cancerhelp.org.uk
Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk
Roy Castle Lung Cancer Foundation ☎ 0333 323 7200 🖱 www.roycastle.org

Colds and influenza

The common cold Mild, acute, self-limiting upper respiratory tract infection more frequent in winter. Most common in children. Adults have on average 2–3/y.

- **Causes** Rhino- (30–50%), corona- (10–15%), influenza (5–15%) viruses most commonly. Others include RSV, entero- and adenoviruses. 25% of colds have no identifiable cause. Infection with ≥2 viruses occurs in 5% of cases
- **Spread** Direct contact and droplet infection
- **Management** Advise patients to rest, take plenty of fluids, and paracetamol or ibuprofen for symptom relief. Usually symptoms resolve in <7d for adults and <14d for younger children. Mild cough may persist for 3wk
- **Complications** Exacerbation of asthma/COPD; secondary infection (bronchitis, pneumonia, conjunctivitis, OM, sinusitis, tonsillitis)

Acute bronchitis Inflammation of major bronchi. Often follows viral URTI especially in winter months. Symptoms include cough ± sputum, breathlessness, and wheeze. On examination, wheeze is often heard without other focal signs. Systemic features may be present, e.g. sweats, fevers, myalgia.

Management Self-limiting illness (settles in <3wk) in normally healthy people. *Consider*: bronchodilators if wheeze is heard; antibiotics—may shorten symptoms but weigh benefits against possible side effects (↑ in community antibiotic resistance and 'medicalizing' a self-limiting condition). If recurrent bronchitis, consider a diagnosis of COPD.

Reasons to prescribe antibiotics immediately[N] Investigate further and/or give antibiotics (e.g. amoxicillin 500mg tds for 5–10d) if:

- Systemically very unwell
- Symptoms/signs of serious illness or complications, e.g. pneumonia
- At high risk of serious complications because of pre-existing co-morbidity, e.g. significant heart, lung, renal, liver, or neuromuscular disease, immunosuppression, CF, or young children born prematurely
- Aged >65y with acute cough and ≥2, or aged >80y with acute cough and ≥1, of the following:
  - Hospitalization in the previous year
  - Type 1 or type 2 DM
  - History of CCF
  - Current use of oral steroids

Influenza Sporadic respiratory illness in winter months causing ~600 deaths/y in non-epidemic years. Minor changes in virus proteins cause annual epidemics. Pandemics occur with major shifts in surface proteins, resulting in global effects as most people lack immunity. *Causes*: Influenza viruses A, B, or C. *Spread*: Droplet infection, person-to-person contact, or contact with contaminated items. *Incubation*: 1–7d.

Presentation Symptoms include fever, cough, sore throat, myalgia, and headache. Usually self-limiting, with acute illness lasting 3–4d. Some symptoms may persist for 1–2wk. Severity varies from asymptomatic to life-threatening complications. Most common complications are secondary bacterial infections, e.g. otitis media, pneumonia, and bronchitis.

Management Rest, fluids, and paracetamol or ibuprofen for fever/symptom control. Treat complications, e.g. antibiotics for chest infection; treatment of exacerbations of COPD or asthma.

Antivirals Oseltamivir and zanamivir are recommended to ↓ complications in adults and children if all of the following apply:

- National surveillance schemes indicate circulating influenza
- The person is in an 'at-risk' group (Box 10.1)
- The person presents with an influenza-like illness and can start treatment within 48h (or within 36h for zanamivir treatment in children) of the onset of symptoms as per licensed indications

Antiviral drugs may also be used for prophylaxis if:

- National surveillance schemes indicate circulating influenza
- An 'at-risk' person has not been immunized and has had close contact with a person with influenza symptoms
- The person can start treatment <48h after contact for oseltamivir (<36h for zanamivir)

Influenza vaccination Offered annually in the UK:

- All patients aged ≥65y—adjuvant vaccine to ↑ effectiveness
- Children aged 2–17y—intranasal live, attenuated vaccine—programme being phased in across the UK
- All aged ≥6mo in high-risk groups (Box 10.1)—quadrivalent, inactivated vaccine <2y; intranasal, live attenuated vaccine if 2–18y; non-adjuvant vaccine aged 18–65y; adjuvant vaccine aged ≥65y

Box 10.1 Risk factors for severe disease with influenza

- Aged ≥65y
- DM
- Immunosuppression
- BMI ≥40kg/m^2
- Pregnant women or ≤2wk postpartum
- Chronic respiratory/heart/renal/liver/neurological disease

High-risk groups eligible for influenza vaccination

- DM
- BMI ≥40kg/m^2
- Chronic respiratory/heart/renal/liver/neurological disease
- Immunocompromised or asplenic patients
- Pregnant women
- Long-stay care facility residents, e.g. nursing homes
- Household contacts of immunocompromised people
- Healthcare and social care workers
- Hajj and Umrah pilgrims

Further information

DH The Green Book. Chapter 19: Influenza. 🖱 www.gov.uk/government/publications/influenza-the-green-book-chapter-19

NICE (2008) Respiratory tract infections (self-limiting): prescribing antibiotics. 🖱 www.nice.org.uk/guidance/cg69

Pneumonia in adults

Acute infection of lung parenchyma. Common condition, affecting 0.5–1% adults annually in the UK. Incidence ↑ with age and peaks in the winter. Mortality rate <1% in those managed in primary care. 22–42% require hospital admission, where mortality rate is 5–14%.

Presentation Acute illness is characterized by:

- Symptoms of an acute lower respiratory tract illness (cough + ≥1 other lower respiratory tract symptom, e.g. purulent sputum, pleurisy, wheeze, pleuritic pain)
- New focal chest signs on examination (consolidation or ↓ air entry, coarse crackles and/or pleural rub)
- ≥1 systemic feature: sweating, fevers, shivers, aches and pains, and/or temperature ≥38°C
- No other explanation for the illness

! The elderly may present atypically, e.g. 'off legs' or acute confusion.

Common causative organisms: Usually bacterial

- *S. pneumoniae* (36%)—➲ p. 630
- *H. influenzae* (10%)—more common among the elderly—➲ p. 632
- *Influenza A* and *B* (8%)—annual epidemics during the winter months; ~3% develop pneumonia—➲ p. 292
- *Mycoplasma pneumoniae* (1.3%)—less common in the elderly. Epidemics occur every 4y in the UK—➲ p. 298
- Gram −ve enteric bacteria (1.3%)
- *C. psittaci* (1.3%)—~20% have history of bird contact—➲ p. 298
- *S. aureus* (0.8%)—more common in the winter months; may be associated with viral infection, e.g. flu—➲ p. 630
- *Legionella* spp. (0.4%)—most common in September/October—>50% related to travel
- No cause identified (45%)

TB ➲ p. 296

Immunocompromised patients ➲ p. 626

Vaccination for prevention

- Influenza ➲ p. 293
- Pneumococcal ➲ p. 630

Differential diagnosis Pneumonitis, e.g. secondary to radiotherapy; malignancy; pulmonary oedema; PE; asthma/COPD exacerbation; TB.

Investigations Often unnecessary in general practice. *Consider*:

- Pulse oximetry Use to assess severity. If oxygen saturation is <94% in air, the patient is hypoxic and requires admission
- CXR If diagnostic uncertainty, symptoms not resolving, or risk of lung cancer. CXR changes may lag behind clinical signs, but should return to normal <6wk after recovery. Persistent changes on CXR >6wk after recovery require further investigation
- Sputum culture If not responding to treatment. If weight ↓, malaise, night sweats, or risk factors for TB (ethnic origin, history of TB exposure, social deprivation or elderly), request mycobacterium culture
- Blood FBC—↑ WCC; ↑ ESR; acute and convalescent titres to confirm 'atypical' pneumonia (*Legionella, C. psittaci, M. pneumoniae*)

Table 10.11 Assessment of severity of pneumonia

⚠ Red flag features	Intermediate features
• Objective evidence of new altered mental state • ↑ respiratory rate: ≥25 breaths/min • New need for O_2 to maintain saturation >92% (or >88% if COPD) • Systolic BP: ≤90mmHg or >40mmHg below normal • ↑ heart rate: >130bpm • Not passed urine in ≥18h (if catheterized, passed <0.5mL/kg/h of urine) • Mottled or ashen appearance • Cyanosis of skin, lips, or tongue • Non-blanching skin rash	• History from patient, friend, or relative of new onset of altered behaviour or mental state • History of acute deterioration of functional ability • ↑ respiratory rate: 21–24 breaths/min • Systolic BP: 91–100mmHg • ↑ heart rate: 91–130bpm (pregnant ♀: 100–130bpm) or new onset arrhythmia • Not passed urine in 12–18h (if catheterized, passed 0.5–1mL/kg/h of urine) • Tympanic temperature <36°C

Management

Consider the need for admission Table 10.11

- **Admit as a blue light emergency**—if any 'red flag' features. If life-threatening infection or considerable delay (>2h), consider administering antibiotics before admission
- **Consider admission**—if any 'intermediate features'. Have a low threshold if: ≥65y, poor social situation, concomitant illness (e.g. heart failure, chronic lung, renal or liver disease, DM, cancer), impairment in immune function (immunosuppressant drugs, steroids), or trauma, surgery, or invasive procedure in the last 6wk

If a decision is made to treat at home

- **Advise not to smoke** Take analgesia, rest, and drink plenty of fluids
- **If no intermediate features** Give antibiotics for 5d, e.g. amoxicillin 500mg tds or doxycycline 200mg on day 1 then 100mg od, or clarithromycin 500mg bd. Review after 3d: if poor response, extend treatment to 7–10d
- **If any intermediate features** Consider dual therapy with amoxicillin 500mg tds + clarithromycin 500mg bd for 7–10d, or monotherapy with doxycycline for 7–10d. Review within 3d: if poor response, admit

Complications Require specialist management—refer.

- Pleural effusion (may be reactive or empyema—pus in the lung cavity)
- Lung abscess (presents with swinging fever and worsening pneumonia)
- Septicaemia—➔ p. 1056
- Metastatic infections
- Respiratory failure
- Jaundice

Further information

NICE (2014) Pneumonia in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg191

NICE (2016, updated 2017) Sepsis: recognition, diagnosis and early management. 🖱 www.nice.org.uk/guidance/ng51

TuberculosisND

Caused by bacteria of the *Mycobacterium tuberculosis* complex. In 2015, there were 10.4 million new cases worldwide and 1.8 million deaths. In the UK, 6240 cases were notified in 2015 and accounted for ~300 deaths.

Risk factors In the UK:

- Born in high-prevalence areas, e.g. India, Pakistan, Nigeria
- Active TB contact in same household; close contacts at school/work
- Previous (especially incomplete) TB treatment—in 2013, 7% of reported TB cases had a diagnosis of TB >12mo previously
- Immunosuppression, e.g. by medications (steroids, chemotherapy) or co-morbidities (HIV, diabetes). People living with HIV accounted for 1.2 million (11%) new cases in 2015 worldwide
- Smoking, alcohol, and drug misuse
- Social factors ~70% of all patients with TB in the UK were resident in the 40% most deprived areas. At particular risk are the homeless, institutionalized people, and people living in overcrowded conditions or prisons

Prevention Bacille Calmette–Guérin (BCG) is a live attenuated strain of bacteria derived from *M. bovis*. BCG vaccination provides immunity lasting ≤15y to 70–80% of recipients. It is given by intradermal injection into the left upper arm. Target groups:

- All infants living in areas where incidence of TB is ≥40/100,000 and infants whose parents or grandparents were born in a country with TB incidence of ≥40/100,000
- Previously unvaccinated new immigrants from countries where there is a high prevalence of TB
- Occupational risk, e.g. healthcare workers, veterinary staff, prison staff
- Contacts of known cases or those living or working in high-prevalence countries for extended periods (generally ≥3mo)

⚠ Do not give other immunizations into the same arm for 3mo.

Screening TB is a notifiable disease. After notification, contact tracing is initiated, usually through chest clinics. Screening may be done with tuberculin test or interferon gamma release assay, depending on the scenario.

Tuberculin skin test Standard in the UK is the Mantoux test. Useful to detect previous exposure to organism (or BCG vaccination) by provoking an immune reaction. A purified protein derivative of *M. tuberculosis* is injected intradermally in the forearm. Response is read after 48–72h (Table 10.12). Test can be suppressed by:

- Hodgkin's disease
- Glandular fever
- Viral infections
- Corticosteroid therapy
- Sarcoidosis
- Live viral vaccines—do not do a tuberculin test <4wk after vaccination
- Immunosuppressant treatment or diseases, including HIV

⚠ If a patient has a +ve tuberculin test—DO NOT give BCG vaccination.

Primary TB Initial infection. Transmitted by droplet infection. A lesion forms (usually pulmonary) which drains to local LNs. Immunity develops and the infection becomes quiescent. May be asymptomatic or include:

- Fever
- Night sweats
- Persistent cough ± sputum
- Haemoptysis
- Pneumonia
- Pleural effusion
- Anorexia
- Weight ↓
- Erythema nodosum

Table 10.12 Tuberculin testing and interpretation of results

Diameter of induration	Positivity	Interpretation
<6mm	Negative	Suggests no TB infection—beware false negatives. Previously unvaccinated people may be given BCG provided there are no contraindications
6–14mm	Positive	Should not be given BCG. May be due to previous TB infection or BCG or exposure to non-tuberculous mycobacteria
≥15mm	Strongly positive	Suggests TB infection or disease. Refer for investigation

Investigations and management

- CXR
- 3× sputum samples for culture and microscopy for acid-fast bacilli
- Refer to a TB specialist service for diagnosis and ongoing management

Post-primary TB Reactivation of a primary infection. Initial lesions (usually in the upper lobes of the lung) progress and fibrose. Other sites may develop disease. Multiple small lesions throughout the body results in miliary TB and is common in immunocompromised patients. Symptoms and signs relate to the organs infected. In all cases, refer for specialist treatment. *Extra-pulmonary disease sites*:

- CNS
- Lymph nodes
- Spine (rarely other bones/joints)
- Peripheral cold abscess
- Pericardium
- Miliary

Treatment^N ❗ Always refer to the chest clinic; 10% are resistant to first-line antibiotics. Specialist management will involve:

- Antibiotic drug treatment with combination regimens—usually 6 months of isoniazid + rifampicin, with pyrazinamide + ethambutol for the first 2 months. All have potentially serious side effects and need monitoring
- Risk assessment for drug-resistant TB and HIV infection. Care coordination and allocation of key worker to monitor treatment concordance, side effects, and clinical response. 'Directly observed therapy' (DOT) is used in high-risk groups (e.g. homeless patients) to ensure concordance. Medications are taken under observation
- Contact tracing

Further information

DH The Green Book. Chapter 32: Tuberculosis. 🖱 www.gov.uk/government/publications/tuberculosis-the-green-book-chapter-32
NICE (2016) Tuberculosis. 🖱 www.nice.org.uk/guidance/ng33
World Health Organization Tuberculosis. 🖱 http://www.who.int/tb/en/

Other respiratory infections

Mycoplasma *Mycoplasma pneumoniae* causes epidemics of lower respiratory tract infection every 3–4y. Spread by droplet infection.

- **Incubation** 12–14d
- **Presentation** Dry, persistent cough ± arthralgia. CXR shows bilateral, patchy consolidation. Infection is confirmed with serology
- **Management** Clarithromycin 500mg bd for 2wk or doxycycline 100–200mg daily for 2wk. Relapse is common. Severe infections may require hospital admission

Respiratory chlamydial infection

- ***C. pneumoniae*** Accounts for 6–19% of community-acquired pneumonia—especially in children/young adults. Clinically indistinguishable from *Mycoplasma pneumoniae*. Treat with doxycycline 100–200mg/d or clarithromycin 500mg bd for 2wk, or azithromycin 500mg od for 3d
- ***C. psittaci*** Infects many animals, but human infection is closely related to contact with birds. Treat as for *C. pneumoniae*

Pertussis (whooping cough)ND Caused by *Bordetella pertussis*.

- **Presentation** Incubation: 7–10d; *Symptoms*:
  - *Catarrhal stage* Symptoms and signs of URTI—lasts 1–2wk
  - *Coughing stage* Increasingly severe, paroxysmal cough with spasms of coughing followed by a 'whoop'—associated with vomiting, cyanosis during coughing spasms ± exhaustion. Lasts 4–6wk, then cough improves over 2–3wk. Chest is clear between coughing bouts
- **Investigation** Microscopy and culture of nasal swabs (special swab and culture medium available from the laboratory); FBC—lymphocytosis
- **Management** Macrolide antibiotics (clarithromycin, erythromycin, or azithromycin) are recommended if onset of cough was in the last 21d
- **Complications** Pneumonia, bronchiectasis, convulsions, subconjunctival haemorrhages, and facial petechiae

Prevention

- **Proven contacts** Treat with clarithromycin or erythromycin
- **Childhood vaccination** ➔ p. 619. Personal or FH of febrile convulsion, FH of epilepsy and well-controlled epilepsy are *not* contraindications—give advice on fever prevention. Defer vaccination if any undiagnosed/evolving neurological condition or poorly controlled epilepsy until the condition is stable—if in doubt, seek specialist paediatric advice
- **Vaccination in pregnancy** Offered from 16–32wk gestation in the UK to ↑ passive immunity in newborn babies. Combination vaccine containing inactivated polio, tetanus, and diphtheria vaccine as well as pertussis is recommended. If missed, can be given until the ♀ goes into labour

❗ Pertussis immunity wanes after vaccination—do not rule out the possibility of pertussis because a person has been vaccinated.

Further information

DH The Green Book. Chapter 24: Pertussis 🖱 www.gov.uk/government/publications/pertussis-the-green-book-chapter-24

Aspergillosis Spores of Aspergillus fungus present in the soil and decaying vegetation can be inhaled any time of the year, but reach peak levels in autumn/winter. Inhaled spores colonize bronchial mucosa and nasal sinuses.

Presentations

- **Asthma** ➔ p. 278
- **Allergic bronchopulmonary aspergillosis** Presents with episodes of eosinophilic pneumonia (characterized by wheeze, cough, fever, and malaise) throughout the year, but worse in late autumn. CXR shows fleeting lung shadows (cleared by expectorating firm, brown plugs of mucus). Untreated → upper lobe fibrosis and 'proximal' bronchiectasis
- **Invasive aspergillosis** Only occurs in the immunocompromised. Aspergillus disseminates from the lung → brain, kidneys, and other organs. Carries very poor prognosis
- **Aspergillus sinusitis** Nasal congestion, headache, and facial pain
- **Aspergilloma** Growth within existing lung cavities (e.g. from previous TB or sarcoidosis). A ball of fungus forms. CXR shows a round lesion with air halo above it. Occasionally results in haemoptysis

Management Refer for specialist management.

Pneumocystis jiroveci (PCP) May be classified as a protozoan or fungus. Causes pneumonia in immunocompromised patients.

- **Presentation** Fever, breathlessness, tachypnoea, dry cough, respiratory failure (± cyanosis)
- **Investigation** CXR normal or 'ground-glass' appearance; sputum culture may be diagnostic
- **Management** If suspected, refer for specialist care. Treatment is with co-trimoxazole or dapsone
- **Prevention** Prophylactic antibiotics (usually co-trimoxazole) are given to AIDS patients with CD4 counts <200 cells/mm^3

Novel corona virus infectionsND *Include:* SARS (sudden acute respiratory syndrome), MERS (Middle East respiratory syndrome) and COVID-19 (corona virus disease 2019). *Spread:* direct contact with infected individual/contaminated surface or aerosol. Incubation: 2–14d (mean 5.5d).

Two stages:

- **Prodrome** Fever (>37.8°C), malaise, headache, myalgia
- **Respiratory phase** Develops after 3–7d—dry cough and breathlessness.

Other symptoms Watery diarrhoea, conjunctivitis, URTI symptoms, e.g. rhinitis, sore throat. Asymptomatic patients may still spread the virus.

>95% recover after 7–10d but 1–2% progress to respiratory failure and/or 2° complications, e.g. pneumonia. If mild symptoms, manage at home—patients must self-isolate for ≥7d or until full recovery. If breathless at rest, peripheral oxygen saturation <92% on air, circulatory compromise, or persistent fever >7d, admit as an acute medical emergency. Personal contacts of patients with suspected/confirmed infections must self-isolate for 14d. A COVID-19 vaccine is likely to be available by 2020/2021.

⚠ Always wear personal protective equipment (PPE) when assessing/managing patients with suspected corona virus infection and pre-warn ambulance/hospital staff if transferring to hospital.

Cystic fibrosis and Kartagener syndrome

Cystic fibrosis (CF) The most common inherited disorder in the UK (prevalence: 1 in 2500). Around 10,800 people in the UK have CF. Median survival has ↑ dramatically and is now >40y, but of the 7500 CF patients in the UK, 6000 are <25y old.

Genetics Results from mutation of a single gene on chromosome 7 (cystic fibrosis transmembrane conductance regulator) essential for salt and water movement across cell membranes. This causes thickened secretions. >2000 different mutations have been described. Autosomal recessive inheritance results in a 1 in 4 chance of having a child with CF if both parents are carriers. ~1 in 25 adults in the UK carries the CF gene. Most common in Caucasians—rare in people of Afro-Caribbean origin.

Screening Several possibilities:
- Pre-conceptual screening Buccal smears to karyotype prospective parents
- Antenatal screening Chorionic villous sampling at ~10wk—for parents with an affected child already or where both parents are +ve on karyotyping
- Neonatal screening ➔ p. 830

Common problems associated with CF Figure 10.4

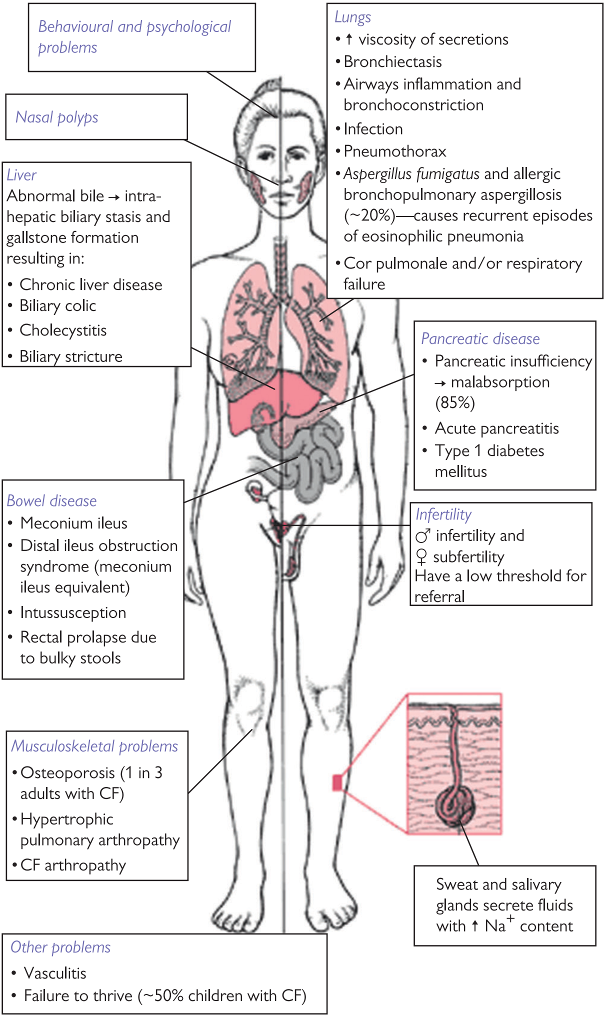
Diagnosis
- Screening
- If clinical suspicion of CF, refer to paediatrics. A +ve sweat test (>60mmol/L on 2 occasions) is diagnostic as is ↑ potential difference across the nasal respiratory epithelium. An elevated sodium level may be found on the sweat test, as well as ↑ chloride:sodium ratio (>1)

Management CF is a multisystem disease requiring a holistic approach to care, which aims to maintain patients' independence, improve quality of life, *and* extend life expectancy. A multidisciplinary team in a specialist CF centre is best placed to achieve this. Patients usually have direct access. Management involves:
- Treatment of lung disease, e.g. with exercise, physiotherapy, antibiotics, and mucolytics
- Maintaining good nutritional state, e.g. pre-meal oral pancreatic enzymes, high-calorie diet, and fat-soluble vitamin supplements (A, D, and E)
- Treatment of complications, e.g. DM, osteoporosis

Further information for patients and professionals

CF Trust ☎ 0300 373 1000 🖱 www.cysticfibrosis.org.uk

Kartagener syndrome (immotile cilia syndrome) Autosomal recessive inherited condition results in abnormal structure and function of cilia. Consists of a combination of bronchiectasis, chronic sinusitis, and male infertility plus situs inversus (transposed heart and abdominal organs). Otitis media and salpingitis are frequent. Specialist treatment is supportive.



Behavioural and psychological problems

Lungs
- ↑ viscosity of secretions
- Bronchiectasis
- Airways inflammation and bronchoconstriction
- Infection
- Pneumothorax
- *Aspergillus fumigatus* and allergic bronchopulmonary aspergillosis (~20%)—causes recurrent episodes of eosinophilic pneumonia
- Cor pulmonale and/or respiratory failure

Nasal polyps

Liver
Abnormal bile → intra-hepatic biliary stasis and gallstone formation resulting in:
- Chronic liver disease
- Biliary colic
- Cholecystitis
- Biliary stricture

Pancreatic disease
- Pancreatic insufficiency → malabsorption (85%)
- Acute pancreatitis
- Type 1 diabetes mellitus

Bowel disease
- Meconium ileus
- Distal ileus obstruction syndrome (meconium ileus equivalent)
- Intussusception
- Rectal prolapse due to bulky stools

Infertility
♂ infertility and
♀ subfertility
Have a low threshold for referral

Musculoskeletal problems
- Osteoporosis (1 in 3 adults with CF)
- Hypertrophic pulmonary arthropathy
- CF arthropathy

Sweat and salivary glands secrete fluids with ↑ Na^+ content

Other problems
- Vasculitis
- Failure to thrive (~50% children with CF)

Figure 10.4 Features of cystic fibrosis

Modified from the *MSD Manual Consumer Version* (Known as the *Merck Manual* in the US and Canada and the *MSD Manual* in the rest of the world), edited by Robert Porter. Copyright 2019 by Merck Sharp & Dohme Corp., a subsidiary of Merck & Co., Inc., Kenilworth, NJ. Available at http://www.msdmanuals.com/consumer. Accessed 20 May 2019.

Interstitial lung disease

Also known as diffuse parenchymal lung disease. Comprises >200 different diseases (many rare) in which inflammation affects the alveolar wall, leading to fluid in the alveolar air spaces.

Presentation Increasing dyspnoea ± cough. More rarely wheeze, pleurisy, and/or haemoptysis. May present incidentally with changes on CXR.

Further assessment

- History of the condition—acute, episodic, chronic?
- Severity—exercise tolerance
- Possible causes:
  - Smoking
  - Hobbies and occupation
  - Usual environment (e.g. lives on a farm) and travel
  - Past medical history (particularly rheumatological symptoms and immunosuppression e.g. HIV) and family history
  - Drugs
- Examine looking for fine inspiratory crackles in the chest, and evidence of systemic disease, e.g. fever, rashes, or other skin changes, eye signs (particularly red eye), hepatomegaly, and/or splenomegaly, arthritis
- Pulse oximetry—↓ peripheral oxygen saturations

Investigations

- **CXR**—diffuse shadowing
- **Urine dipstick**—for protein and blood
- **Blood**—FBC, ESR, liver and kidney function tests, thyroid function tests, autoimmune profile
- **Lung function tests**—usually show restrictive picture—rarely no abnormalities or obstructive picture

Classification Table 10.13

Hypersensitivity pneumonitis Also known as extrinsic allergic alveolitis, farmer's lung, and bird fancier's lung. Inhaled particles (e.g. fungal spores, avian proteins) cause an allergic reaction in lungs of hypersensitive individuals. May present as an acute or chronic reaction, or both may occur together.

- **Acute reaction** 2–4h post exposure. Fever, malaise, dry cough, shortness of breath
- **Chronic reaction** Malaise, weight ↓, exertional dyspnoea, fine crepitations in both lung fields

Investigations

- **Blood** FBC: ↑ neutrophils (acute reaction); ESR ↑ (acute reaction)
- **CXR** May be normal or show typical changes (shadowing, widespread small nodules or ground glass appearance)
- **Diagnosis** Based on history and high-resolution CT scan findings. Serum precipitins to the provoking factor are found in ≥90%

Management If possible prevent further exposure to the allergen. In all cases refer for specialist advice. Treatment is usually with corticosteroids. If occupational exposure, may qualify as industrial disease and be eligible for compensation—➔ p. 90.

Table 10.13 Classification of interstitial lung disease

Classification	Causes
Acute	Infective: • Bacterial (TB) • Viral (chicken pox, measles) • Fungal Allergy—drugs, fungi, helminths Toxins—drugs, gases Haemodynamic—LVF, fluid overload, renal failure Vasculitis Adult respiratory distress syndrome
Episodic	Eosinophilic pneumonia, e.g. allergic bronchopulmonary aspergillosis Vasculitis, e.g. eosinophilic granulomatosis with polyangiitis (formerly Churg–Strauss syndrome) Hypersensitivity pneumonitis Cryptogenic organizing pneumonia
Chronic due to occupational or environmental exposure	Dust-induced (➔ p. 306)—asbestosis, silicosis, coal worker's pneumoconiosis, siderosis (iron) Farmer's lung Bird fancier's lung Radiation Drugs, e.g. nitrofurantoin, sulfasalazine, gold, penicillamine, aspirin, amiodarone, bleomycin, methotrexate, hydralazine, heroin, methadone, oxygen
Chronic with evidence of systemic disease	Connective tissue disease, e.g. RA, Sjögren's syndrome, SLE Neoplastic, e.g. lymphoma Vasculitis, e.g. granulomatosis with polyangiitis (formerly Wegener's granulomatosis), Goodpasture's syndrome Sarcoidosis Inherited disorders, e.g. tuberous sclerosis, neurofibromatosis Miscellaneous, e.g. HIV, inflammatory bowel disease, post bone marrow transplant, amyloidosis
Chronic without evidence of systemic disease	Idiopathic pulmonary fibrosis Chronic aspiration

⚠ Advise all patients with interstitial lung disease to stop smoking. This results in better prognosis for their interstitial lung disease. Furthermore, patients with chronic interstitial lung disease are at substantially ↑ risk of lung cancer.

Idiopathic pulmonary fibrosis (IPF) 6000 people are diagnosed every year in the UK. ♂ > ♀. Incidence ↑ with age. ~85% diagnoses made in people >70y. Progressive condition of unknown cause with insidious onset. Can only be diagnosed if other causes of interstitial lung disease have been excluded and symptoms present >3mo.

Risk factors

- Smoking
- Environmental exposure, e.g. metals dusts, animal dusts
- Chronic viral infection, e.g. EBV, hepatitis C
- GORD

Presentation

- Progressive exertional dyspnoea
- Dry cough
- Clubbing (>50%)
- Fine 'Velcro-like' crepitations
- Malaise
- Weight ↓
- Central cyanosis and right heart failure (advanced cases)

Investigations

- **CXR** Diffuse shadowing (although may be normal)
- **Lung function tests** Restrictive picture

Differential diagnosis

- LVF
- COPD
- Inhalant exposure (O_2, NO_2)
- Radiation
- Other causes of lung fibrosis—dust exposure (coal, asbestos, silica, farmer's lung, bird fancier's lung)

Management Refer to a respiratory physician for diagnosis and advice on management. Treatment, where appropriate, is with oral steroids + azathioprine. Pulmonary rehabilitation may be helpful. Lung transplant is a last option. Most patients have poor prognosis with median survival 3y. A subgroup with fibrotic non-specific interstitial pneumonia (NSIP) has substantially better prognosis with >50% surviving 5y.

❗ Patients with IPF have a 10× ↑ risk of lung cancer. This risk is multiplicative with that from smoking. Patients with IPF who smoke 20 cigarettes a day may have a 200× ↑ risk of lung cancer compared with non-smokers without IPF.

Sarcoidosis Multisystem inflammatory disease of unknown cause characterized by non-caseating granuloma. Annual incidence in the UK is 7/100,000. Typically presents with lung granuloma in a young adult. ♀ > ♂.

Non-respiratory manifestations of sarcoidosis

- Fever and malaise
- Erythema nodosum
- Lupus pernio (blue-red nodules on the nose, face, and/or hands)
- Scar infiltration
- Enlarged lacrimal glands
- Hypopyon
- Uveitis
- Arthralgia
- Hepatosplenomegaly
- Arrhythmias
- Heart failure
- Pericardial effusion
- Cranial and/or peripheral nerve palsies
- Seizures
- Hypercalcaemia
- Renal stones
- Lymphadenopathy

Acute sarcoidosis (Löfgren's syndrome)

- Polyarthralgia
- Swinging fever
- Erythema nodosum
- Bilateral hilar lymphadenopathy on CXR

Insidious onset CXR shows hilar lymphadenopathy—incidental finding in 30–50%. If symptomatic, usually presents with tiredness, malaise, weight ↓, and/or arthralgia. 15% have lung symptoms with gradual onset of progressive exertional dyspnoea and dry cough.

Management Refer any patient with bilateral hilar lymphadenopathy for further investigation. For patients with confirmed sarcoidosis, specialist management is needed. Steroids are the first-line treatment, but should only be used if:

- Progressive disease (on imaging or lung function testing)
- Significant symptoms
- Extra-pulmonary disease requiring treatment

Rarely, if steroids are not controlling disease progression or symptoms, methotrexate will be added. Inhaled steroids may be helpful to control cough, but do not influence disease progression. For patients with severe symptoms, pulmonary rehabilitation may be helpful. Lung transplant may be considered for patients with end-stage pulmonary sarcoidosis.

Prognosis Variable natural history, so predicting the course of disease and prognosis is difficult. Spontaneous remission occurs in 55–90% of patients with stage I radiological disease, 40–70% with stage II disease, and 10–20% with stage III disease. Most remissions occur in the first 6mo. Mortality ranges from 1–5%, due to pulmonary, myocardial, or CNS involvement.

Occupational lung disease ➜ p. 306

Further information

British Thoracic Society Interstitial lung disease guideline. *Thorax* **63**:v1–v58. 🖱 https://www.brit-thoracic.org.uk/document-library/clinical-information/interstitial-lung-disease/ild-guidelines/bts-interstitial-lung-disease-guideline/

NICE (2013, updated 2017) Idiopathic pulmonary fibrosis in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg163

Patient support

British Lung Foundation ☎ 03000 030 555 🖱 www.blf.org.uk

Occupational lung disease

Exposure to gases, vapours, and dusts at work can lead to lung disease.

Coal-worker's pneumoconiosis 90% of all compensated industrial lung disease in the UK. 'Pneumoconiosis' means accumulation of dust in the lungs and tissue reaction to its presence. Incidence is related to total dust exposure. Divides into:

- **Simple pneumoconiosis** Deposition of coal dust in the lung. Graded on CXR appearance. Grading determines whether disability benefit is payable in the UK. Effect on lung function is debated. Predisposes to progressive massive fibrosis
- **Progressive massive fibrosis** Round fibrotic masses several cm diameter form in the upper lobes. Presents with exertional dyspnoea, cough, black sputum, and eventually respiratory failure. Symptoms progress (or even start) after exposure to coal dust has ceased. Lung function tests show a mixed restrictive and obstructive picture with loss of lung volume, irreversible airflow limitation, and ↓ gas transfer

Asbestosis Before legislation banning its use, exposure was widespread and occurred particularly in naval shipyards and power stations. Effects of asbestos exposure—Table 10.14. Consider diagnosis in relatives who came into contact with asbestos while washing clothes etc. too—they can claim compensation if affected.

Silicosis Uncommon. Affects stonemasons, pottery workers, workers exposed to sand-blasting, and fettlers (remove sand from metal casts). Caused by inhalation of silica. CXR appearance is distinctive. Presents with exertional dyspnoea ± cough.

Lung function tests as for progressive massive fibrosis. Associated with ↑ risk of lung cancer and TB.

Byssinosis Affects cotton mill workers. Symptoms (tightness in the chest, cough, and breathlessness) start on the 1st day back at work after a break (Monday sickness) with improvement as the week progresses. CXR is normal.

Berylliosis Rare. Long latent period. Affects workers in the aerospace, nuclear power, and electrical industries and their close relatives. Presents similarly to sarcoidosis (➔ p. 304).

Iron (siderosis), barium (baritosis) and tin (stannosis) dust inhalation Result in dramatic dense nodular shadowing on the CXR, but effects on lung function and symptoms are often minimal.

Occupational asthma >200 industrial materials cause occupational asthma. Accounts for 17% of all adult asthma cases. Important causes are recognized occupational diseases in the UK—patients may be eligible for statutory compensation if they apply <10y after leaving the occupation in which asthma developed. Suspect if a patient has symptoms which improve on days away from work/holiday.

Hypersensitivity pneumonitis ('farmer's lung') ➔ p. 302

Management In all cases refer to a respiratory physician for confirmation of diagnosis (essential if seeking compensation) and advice on management.

Table 10.14 Conditions caused by asbestos exposure

Condition	Asbestos exposure	Features/management
Benign pleural effusion	Usually occurs <20y after exposure	Increasing dyspnoea ± pleuritic pain Refer for drainage of effusion. May be recurrent and require pleurodesis
*Bilateral diffuse pleural thickening**	Follows light or moderate exposure to asbestos. May progress even in the absence of further exposure	Defined as pleural thickening >5mm thick covering >¼ of the chest wall Symptoms: exertional dyspnoea Lung function tests: restrictive picture Treatment is symptomatic
*Asbestosis**	Follows heavy exposure after a 5–10y interval	Presents with progressive dyspnoea, finger clubbing, and basal end-expiratory crackles CXR: 'honeycomb lung'—diffuse streaky shadowing Lung function tests: severe restrictive defect and ↓ gas transfer Treatment is symptomatic
*Mesothelioma**	Can follow even light exposure to asbestos. 20–40y time lag between exposure and appearance of disease	Presents with ↑ shortness of breath ± pleuritic pain. Examination and CXR reveal unilateral (rarely bilateral) effusion There is no effective active treatment. Palliative care—➔ p. 1011 Median survival is 2y from diagnosis
*Asbestos-related lung cancer**	Patients exposed to asbestos who have evidence of that exposure (pleural plaques, bilateral pleural thickening, or asbestosis) have an ↑ risk of bronchial carcinoma—usually adenocarcinoma. Smokers exposed to asbestos have a 5× ↑ risk compared to non-smokers exposed to asbestos Manage as for lung cancer—➔ p. 290	

* Eligible for industrial injuries benefit in the UK.

Benefits ➔ p. 91

Notification and compensation ➔ p. 90

Further information

European Lung White Book 🖱 www.erswhitebook.org

Patient support

British Lung Foundation ☎ 03000 030 555 🖱 www.blf.org.uk

Snoring and obstructive sleep apnoea

Snoring During sleep, the pharyngeal airway narrows due to ↓ dilator muscle tone. Snoring is vibratory noise generated from the pharynx and soft palate as the air passes through this narrowed space. Further narrowing produces louder snoring, laboured inspiration, and eventually apnoeic episodes. Social consequences are the usual reason for the patient to seek help. They can be distressing: banishment from the bedroom, marital disharmony, no holidays, fear of travelling or falling asleep in a public place, etc.

❗ Snoring may be used by the spouse as an excuse to leave the marital bed and may actually be trivial/absent. If suspected, ask the patient to bring a recording of the offending noise.

Obstructive sleep apnoea Occurs when the pharyngeal airway completely closes during sleep resulting in apnoeic episodes. ↑ inspiratory effort is sensed by the brain and a transient arousal provoked. A few of these arousals do not matter, but many (sometimes hundreds) per night → fragmented sleep and consequent daytime sleepiness. Affects 4% ♂ and 2% ♀ in the UK.

Clinical features

- **Dominant features** Excessive daytime sleepiness (not tiredness—Epworth Sleepiness Scale is a useful assessment tool), impaired concentration, snoring
- **Other features** Unrefreshing sleep, choking episodes during sleep, witnessed apnoeic episodes, restless sleep, irritability/personality change, nocturia, ↓ libido

Causes of snoring and sleep apnoea Overweight (neck circumference >43cm), nasal congestion, evening alcohol/sedatives, large tonsils, receding lower jaw, smoking, hypothyroidism, menopause.

Management

Snoring without sleep apnoea

- **Initial approaches** Suggest changing sleeping position (discourage from sleeping on back); elevate head of the bed (e.g. prop up on bricks—can ↓ nasal congestion); limit number of pillows to 1 thick/2 thin pillows to maximize pharyngeal size; ↓ weight if obese; ↓ or stop evening alcohol/sleeping tablets; suggest partner tries ear plugs (purchase from chemist—takes several nights to get used to wearing them)
- **If clinically indicated**
  - Nasal congestion—start beclometasone nasal spray (applied head downwards) 2 puffs bd ± ipratropium bromide nasal spray 2 puffs nocte
  - Check TFTs to exclude hypothyroidism
- **If simple measures fail** Refer to:
  - Dentist or ENT for a mandibular advancement device
  - ENT for surgery—septal straightening, polypectomy, turbinate reduction, tonsillectomy or uvulopalatopharyngoplasty

Sleep apnoea

- Advise patients to: ↓ weight if obese; ↓ or stop evening alcohol/sleeping tablets *and*

- Refer to a sleep unit or physician with a special interest in sleep problems. If diagnosis is proven and causing significant daytime sleepiness, usual treatment is with CPAP therapy at night. Mandibular advancement devices are alternatives for patients who cannot tolerate CPAP or have very mild symptoms with no daytime sleepiness. Occasionally, if large tonsils, referral to ENT for surgery is warranted
- Complications: ↑ BP, DM, ↑ risk of stroke and road traffic accidents

The Epworth Sleepiness Scale How likely are you to doze off or fall asleep in the following situations, in contrast to feeling just tired?

This refers to your usual way of life in recent times. Even if you have not done some of these things recently, try to work out how they would have affected you.

Situation	Chance of dozing
Sitting and reading	☐
Watching TV	☐
Sitting inactive in a public place (e.g. a theatre or a meeting)	☐
As a passenger in a car for an hour without a break	☐
Lying down to rest in the afternoon when circumstances permit	☐
Sitting and talking to someone	☐
Sitting quietly after a lunch without alcohol	☐
In a car, while stopped for a few minutes in traffic	☐

0 = no chance of dozing
1 = slight chance of dozing
2 = moderate chance of dozing
3 = high chance of dozing

If score >10—consider sleep apnoea.

Murray W. Johns. A New Method for Measuring Daytime Sleepiness: The Epworth Sleepiness Scale. *Sleep* (1991) 14 (6): 540–545, doi: 10.1093/sleep/14.6.540. Translated and reproduced by permission of Oxford University Press on behalf of the Sleep Research Society.

⚠ **Driving** Warn patients NOT to drive if sleepy. Once diagnosis is confirmed, must inform DVLA and insurance company (➲ p. 98).

Sleep apnoea in children Common in children aged 2–7y in association with tonsil enlargement during URTI. Sleep disruption can cause daytime sleepiness, hyperactivity, poor attention span and bad behaviour.

If tonsils are big enough to produce sleep apnoea in the absence of current infection, refer to ENT for consideration of tonsillectomy.

Patient support

The Sleep Apnoea Trust (SATA) 🖱 www.sleep-apnoea-trust.org

Chapter 11

Endocrinology

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Symptoms of endocrine disease

Hormones secreted by the endocrine system perform a wide range of functions. Therefore, clinical presentation of different endocrine disorders varies widely from non-specific symptoms such as tiredness, to very specific signs such as delayed puberty. Specific features depend on the gland and hormones involved.

Polydipsia Over-frequent drinking of fluid—often associated for logical reasons with polyuria. Ask if associated with thirst. Take a history of fluid intake. If no history of excess fluid intake and capillary blood glucose/fasting blood glucose/HbA1c is normal, investigate further with U&E, Cr, eGFR, and Ca^{2+}.

Common causes

- Change in lifestyle: diet/activity/exercise level—may be associated with polyuria but no other symptoms. No history of thirst
- DM—usually accompanied by a history of thirst

Other causes Diarrhoea, diabetes insipidus (➲ p. 341), ↑ Ca^{2+} (➲ p. 336), compulsive water drinking (may be a feature of psychotic illness), phosphorus poisoning.

Polyuria Passage of excessive urine. Check the patient does not mean frequency of urination. It can be difficult to distinguish the two. Causes are similar to those of polydipsia and the 2 symptoms are related. Take a history of fluid intake. If no history of excess fluid intake and capillary blood glucose/fasting blood glucose/HbA1c is normal, investigate further with MSU (for M,C&S), U&E, Cr, eGFR, and Ca^{2+}.

Consider

- DM
- Diabetes insipidus
- Hypercalcaemia
- Excessive intake—due to change in lifestyle or psychiatric conditions, e.g. schizophrenia
- Chronic renal failure
- Drugs—diuretics, caffeine, alcohol

Glycosuria Often detected incidentally on urine dipstick. *Causes*:

- DM
- Pregnancy
- Sepsis
- Renal tubular damage
- Low renal threshold

In all cases check HbA1c/fasting blood glucose (+ glucose tolerance test if pregnant). Check immediate capillary blood glucose if other symptoms suggestive of DM.

Hirsutism Affects 10% of ♀. Excess hair in androgenic distribution. *Causes*:

- Most cases are idiopathic; there may be a family history
- Drugs—phenytoin; corticosteroids; ciclosporin; androgenic oral contraception; anabolic steroids; minoxidil; diazoxide
- Polycystic ovarian syndrome (PCOS)
- Late-onset congenital adrenal hyperplasia (rare)
- Cushing's syndrome
- Ovarian tumours (rare)

Assessment

- **History** Long-standing or recent onset, family history, ethnic origin (more common if of Mediterranean origin), menstrual history
- **Examination** Distribution of excess hair

Investigation Women with longstanding hirsutism (since puberty) and regular periods need no further investigation, unless abnormal signs. *Otherwise*: blood—testosterone (↑ in PCOS, androgen-secreting tumour, late-onset congenital adrenal hyperplasia); LH/FSH ratio (>3:1 suggests PCOS).

Refer to gynaecologist or endocrinologist if Recent onset, abnormal blood tests, virilism, galactorrhoea, menstrual disturbance, infertility, and/or pelvic mass.

Treatment of idiopathic hirsutism

- Cosmetic—bleaching, shaving, waxing, depilatory creams, electrolysis
- Weight ↓ in obese individuals
- Psychological support
- Topical eflornithine ↓ growth of unwanted facial hair. Continuous use for >8wk is required before benefit is seen. Must be used indefinitely to prevent regrowth. Discontinue if no improvement in 4mo
- Oral medication—all must be taken for ≥6mo to take effect and none abolish the problem. In all cases continue treatment until acceptable level of hair growth then stop. Relapse usually follows withdrawal and repeat courses are then required. *Drugs used*: COC pill containing desogestrel or co-cyprindiol; spironolactone

Sweating ➔ p. 575

Facial flushing ➔ p. 568

Delayed or precocious puberty ➔ p. 871

Obesity ➔ p. 152

Metabolic syndrome (syndrome X; insulin resistance syndrome) Impaired glucose tolerance or DM, insulin resistance (in patients on insulin, suggested by insulin doses >1 unit/kg/d) + other risk factors for CVD including:

- Truncal obesity—waist circumference >0.9m (♀); >1.0m (♂)—subtract 0.1m from these figures for people of South Asian extraction
- ↑ BP >135/80mmHg
- Dyslipidaemia—serum HDL <1.2.mmol/L (♀) or <1.0 (♂); fasting serum triglycerides >1.8mmol/L

Associated with high risk of CVD. Treat risk factors aggressively.

Tiredness and lethargy ➔ p. 502

Diabetes mellitus

Diabetes mellitus (DM) is a common syndrome caused by lack, or ↓ effectiveness, of endogenous insulin. It affects 3% of the UK population and is characterized by ↑ blood glucose + abnormalities of carbohydrate/lipid metabolism.

Classification of primary diabetes

Type 1 Occurs at any age but more common in those aged <30y. Autoimmune disease—islet cell antibodies may be present initially. Associated with other autoimmune disease and certain genotypes (HLA DR3/4—although identical twin concordance ≈30%.). Patients are prone to profound weight ↓ and ketoacidosis. Insulin is needed from diagnosis.

❗ Do not measure C-peptide and/or diabetes-specific autoantibody titres to confirm type 1 DM in adults.

Type 2 80–90% patients with DM. ♂:♀ ≈3:2. Prevalence is rising. Progressive disease resulting from ↓ insulin secretion *and* insulin resistance. Onset is often insidious; ½ have complications at diagnosis. *Risk factors*:

- Age >65y
- Obesity
- Pre-diabetes
- FH of DM (identical twin concordance ≈100%)
- Ethnic group—South Asians/Afro-Caribbeans have 5–10× ↑ risk
- PMH of gestational diabetes or a baby >4kg at birth

Latent autoimmune diabetes in adulthood (LADA) 6–10% of patients with type 2 DM. Characterized by antiglutamic acid decarboxylase (GAD) antibodies. Associated with higher risk of ketoacidosis and ↑ risk of progression to insulin dependence. Suspect if type 2 DM and:

- Absence of metabolic syndrome features
- Uncontrolled hyperglycaemia despite oral agents and/or
- Other autoimmune diseases (e.g. thyroid disease, pernicious anaemia)

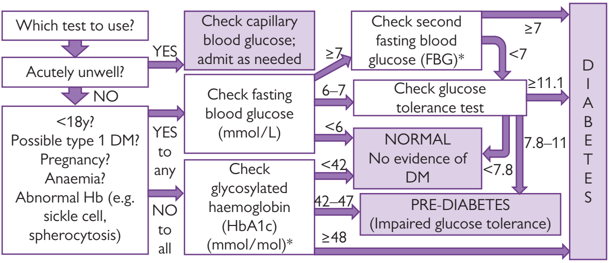
Maturity-onset diabetes of the young (MODY) 1–2% of patients with DM. Present <25y and there is a family history. Genetic syndrome with autosomal dominant inheritance. Gene mutations involved: HNF1-α (70%); HNF1-β; HNF4-α; glucokinase. Gene testing is important to identify the type of MODY, as treatment differs according to type.

Other (secondary) causes of DM

- **Drugs** Steroids, thiazides
- **Pancreatic disease** Pancreatitis, surgery, pancreatic cancer, haemochromatosis, cystic fibrosis
- **Endocrine disease** Cushing's disease, acromegaly, thyrotoxicosis, phaeochromocytoma, pregnancy
- **Others** Glycogen storage diseases, insulin receptor antibodies

⚠ Blood glucose may be temporarily ↑ during acute illness, after trauma or surgery, or during short courses of blood glucose-raising drugs (see 2° causes). If HbA1c ≥48mmol/mol DM is likely.

Testing for diabetes Figure 11.1 ❗ Consider referral for an urgent (<2wk) direct access CT scan (or USS if CT is not available) to assess for pancreatic cancer if aged ≥60y, weight ↓, and new-onset DM[N].


Which test to use?
Acutely unwell?
YES
NO
Check capillary blood glucose; admit as needed
<18y? Possible type 1 DM? Pregnancy? Anaemia? Abnormal Hb (e.g. sickle cell, spherocytosis)
YES to any
NO to all
Check fasting blood glucose (mmol/L)
Check glycosylated haemoglobin (HbA1c) (mmol/mol)*
≥7
6–7
<6
<42
42–47
≥48
Check second fasting blood glucose (FBG)*
≥7
<7
Check glucose tolerance test
≥11.1
7.8–11
<7.8
NORMAL No evidence of DM
PRE-DIABETES (Impaired glucose tolerance)
DIABETES

* For patients with symptoms (polyuria, polydipsia, weight ↓), one FBG is sufficient. If no symptoms, confirm FBG with a second sample; consider confirming an abnormal HbA1c with a second sample.

Figure 11.1 Diagnosis of diabetes

Presentation

- **Acute** Ketoacidosis or hyperosmolar non-ketotic coma (➲ p. 1083)
- **Sub-acute** Weight ↓, polydipsia, polyuria, lethargy, irritability, infections (candidiasis, skin infection, recurrent infections slow to clear), genital itching, blurred vision, tingling in hands/feet
- **With complications** Skin changes (➲ p. 327), neuropathy (➲ p. 329), nephropathy (➲ p. 325), arterial (➲ p. 326) or eye disease (➲ p. 328)
- **Asymptomatic** Incidental finding or through risk stratification

Risk stratification[N] Use a risk stratification tool (e.g. Diabetes Risk Score or QDiabetes) to assess all patients >40y, or >25y if of South Asian, Chinese, Afro-Caribbean, or black African origin, from hard-to-reach populations (e.g. homeless) or with other medical conditions that predispose to DM (e.g. pancreatitis).

- **If low/intermediate risk** Give lifestyle advice and reassess in 5y
- **If high risk** Or of South Asian/Chinese ethnic origin and BMI >23kg/m^2, check FBG or HbA1c. Management—Table 11.1

Pre-diabetes (non-diabetic hyperglycaemia) FBG 6.1–6.9mmol/L or HbA1c 42–47mmol/mol. Risk factor for DM and CVD. Follow-up with annual FBG or HbA1c. 4%/y develop DM. Offer lifestyle advice aiming to normalize blood sugar—a 5% weight ↓ results in ~80% ↓ risk of progression to DM over 3y. Treat CVD risk factors aggressively.

Diabetes and pregnancy ➲ p. 806

Further information

NICE (2011) Preventing type 2 diabetes: risk identification and interventions for individuals at high risk. 🔗 www.nice.org.uk/guidance/ph35

Table 11.1 Diabetes risk stratification for high-risk patients

FBG (mmol/L)	HbA1c (mmol/mol)	Action
<5.5	<42	Provide lifestyle advice; reassess every 3y
5.5–6.9	42–47	Treat as for pre-diabetes
≥7	≥48	Consider type 2 DM

Organization and monitoring of care

Aims of diabetic care

- Alleviation of symptoms
- Minimization of complications
- ↓ in early mortality
- Quality of life enhancement
- Education of the patient and family/carers

Features of well-organized care

- Use of a register and structured records (available as part of in-house computer software)
- 6-monthly review with recall system and follow-up of defaulters
- Protocol for patient-centred care including provision of personal care plans tailored to each individual and including self-management plans
- Provision of protected time for the clinic
- Availability of good-quality written information for patients
- Open access for patients to receive advice
- Multidisciplinary team covering all aspects of diabetes care—e.g. GPs, diabetes nurse specialists/assistants, educators, dieticians, podiatrists
- Quality monitoring through audit and patient feedback
- Continuing education for professional staff

Routine diabetic review Each patient with DM requires 6-monthly review (or more frequent as necessary). This should include a thorough annual review of all aspects of disease and care. Reviews should cover:

- **Problems** Recent life-events; new symptoms; difficulties with management since last visit
- **Review of:**
  - Indices of control, e.g. HbA1c
  - Self-monitored results and discussion of their meaning
  - Lifestyle—dietary behaviours; physical activity; smoking
  - Diabetes education—including referral to a structured education programme for those newly diagnosed and information on lifestyle, self-care, support, and when to step up treatment/seek further medical help
  - Skills, e.g. injection technique for patients on insulin
  - Foot care
  - Review of blood glucose, lipid and BP therapy, and results
  - Other medical conditions and therapy affecting DM
  - Immunizations—influenza ± pneumococcal vaccination
  - Depression screening (➔ p. 173)
- **Review of complications** Annual review—more frequent if established complications. Cardiovascular disease; nephropathy; neuropathy; eye disease; foot problems; erectile dysfunction
- **Review of services** Annual review—more frequent if problems
- **Analysis and planning** Agreement on the main points covered, targets for coming months, changes in therapy, interval to next review
- **Recording** Completion of structured record ± patient-held record

❗ 7–10% of patients in long-term residential care have DM. Patients in residential care with DM tend to be neglected. Agree a diabetes care plan for each affected resident and ensure at least annual diabetic review.

Monitoring blood glucose All patients can achieve good levels of control (Table 11.2). Poorer control is acceptable in the elderly or others with limited life expectancy as long as they are symptom free.

- **Finger-prick capillary glucose monitoring** Essential for all patients using insulin or who are pregnant. In addition, offer to people with type 2 DM if evidence of hypoglycaemic episodes or on oral medication that may ↑ risk of hypoglycaemia while driving or operating machinery
  - Explain the range of suitable monitoring devices available and train in the use of the selected method
  - Frequency of self-monitoring varies according to need
  - Set targets for preprandial glucose levels
  - Assess skills (and meters) yearly or if problems self-monitoring
  - Evaluate reliability of results by comparison with HbA1c results and results obtained at review
- **Glycosylated haemoglobin (HbA1c)** Measure at least 2×/y. Represents average blood glucose control over the previous 6–8wk

❗ Patients with type 2 DM who are taking sulfonylureas or glinides must check blood glucose regularly when driving, so will need blood glucose meters and a supply of testing sticks.

Table 11.2 Indices of control

Measure	Target
Capillary blood glucose (mmol/L)	Fasting blood glucose on waking 5–7 Pre-meals 4–7; >90min after eating 5–7 Before driving ≥5
Urine	−ve (postprandial sugars <0.5%)
HbA1c[a] (normal 20–42mmol/mol)—measure every 2–6mo depending on control	Type 2 DM and: • Drug causing hypoglycaemia ≤53mmol/mol • Diet controlled or drug not causing hypoglycaemia ≤48mmol/mol Type 1 DM ≤48mmol/mol
Serum cholesterol	Aim to ↓ non-HDL cholesterol by >40% from baseline
BMI (kg/m^2)	25–30
BP (mmHg)	<140/80—uncomplicated type 2 DM <135/85—uncomplicated type 1 DM <130/80—if any renal, foot, eye, or cardiovascular complications of type 1 or type 2 DM

[a] Adjust target to the individual; if very elderly or limited life expectancy, relax HbA1c aiming just to keep the patient asymptomatic.

Further information

NICE (2015, updated 2016) Type 1 diabetes in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng17

NICE (2015, updated 2017) Type 2 diabetes in adults: management. 🖱 www.nice.org.uk/guidance/ng28

Patient advice and support

Diabetes UK ☎ 0345 123 2399 🖱 www.diabetes.org.uk

Management of diabetes: education

Education is an essential aspect of diabetic care. Diabetes is a chronic condition, and however well it is managed in the clinic, the patients must manage their own disease the rest of the time. Everyone with DM should receive education through a structured, quality-controlled education programme, e.g. diabetes education and self-management for ongoing and newly diagnosed (DESMOND). Education enables patients and their carers to become equal partners in the management of their disease.

General knowledge Information about:

- DM, its progressive nature, complications, and aims of management
- Structure of diabetic services and ways to access them
- Equipment required and usage instructions—syringes, needles, blood testing equipment, etc.
- Free prescriptions if requiring drugs or insulin to control diabetes
- Problems of pregnancy (♀ of childbearing age only)
- Alert bracelets/tags—Medic-Alert (☎ 01908 951045 🖱 www.medicalert.org.uk) or Medi-Tag (☎ 0121 200 1616 🖱 www.medi-tag.co.uk) provide engraved jewellery, watches, and tags

Diet Patients do *not* need a separate diet from the rest of the family or expensive 'diabetes' food products. A diabetic diet is a healthy diet.

- Aim for ≥50% of calorie intake from fibre-rich carbohydrate, with minimum fat (especially saturated), refined carbohydrate, and alcohol
- Adjust total calorie intake according to desired BMI. If overweight, aim for initial 5–10% weight loss. Consider referral for bariatric surgery if referral criteria are met (➔ p. 153)
- Recommend ≥5 portions of fresh fruit or vegetables/d
- Spread food intake evenly across the day
- Diet sheets are available from Diabetes UK and should be provided
- Ready-made meals, processed foods, and alcohol are often sources of hidden sugar

Immunizations Offer annual influenza and single-dose pneumococcal vaccine to all patients with diabetes.

Psychological problems Discuss concerns about DM. Arrange counselling/refer to self-help resources as needed. Teenagers with diabetes can be a particularly difficult group to manage. Often control is poor due to a combination of rapid bodily changes and rebellion against the diagnosis of DM. Support information and advice given in specialist clinics.

Exercise Encourage regular exercise.

- Review activity at work, and in getting to and from the workplace, hobbies, and physical activity in the home
- Advise physical activity can ↑ insulin sensitivity, ↓ BP, and improve blood lipid control
- If appropriate, suggest regular physical activity tailored to individual ability (e.g. brisk walking for 30 min/d; exercise prescription)

Smoking Advice on and assistance with smoking cessation (➔ p. 156).

Foot care ➔ p. 330

Driving

Group 2 licence (bus and lorry)

- **On insulin and/or sulfonylurea or glinide medication** Inform the DVLA. 1y licence issued if full awareness of hypoglycaemia; no episodes of severe hypoglycaemia in the past 12mo; monitoring capillary blood glucose ≥2×/d regularly (even when not driving), ≤2h before starting a journey, and every 2h while driving. Must use a blood glucose meter with >3mo memory
- **Other antidiabetic medication** Must notify the DVLA but can drive
- **Diet controlled** No need to inform the DVLA

Group 1 licence (car and motorcycle)

- **Insulin controlled** Inform the DVLA. May drive if adequate awareness of hypoglycaemia; ≤1 episode of severe hypoglycaemia while awake in the past 12mo and last episode >3mo ago; capillary blood glucose monitoring ≤2h before a journey and every 2h whilst driving
- **Sulfonylurea or glinide medication** No need to inform the DVLA. Must be under regular medical review. May drive if adequate awareness of hypoglycaemia; ≤1 episode of severe hypoglycaemia while awake in the past 12mo and last episode >3mo ago. Capillary glucose monitoring as for insulin if needed clinically—usually if new medication (<3mo), if any episodes of hypoglycaemia and/or ↓ awareness of hypoglycaemia
- **Diet controlled or other antidiabetic medication** No need to inform the DVLA

❗ Drivers should also inform their insurance company. Other conditions associated with DM (e.g. visual impairment, CHD) may preclude driving.

Employment Advise those on insulin that certain jobs are no longer possible, e.g. working on scaffolding or with dangerous machinery; joining the police or armed services. Jobs without these hazards should pose no problems although the patient might wish to tell his/her employer. Special advice may be needed for shift work.

Travel Be aware of insurers catering for diabetic travellers. *Give advice on*: management of change in time zones (no change needed with oral medication or if taking insulin and <4h time difference); transport and storage of insulin (needs refrigeration in hot climates); keeping monitoring/injection equipment in hand-luggage; differences in insulin types and concentrations between countries; travel-related illness (especially gastroenteritis); need for immunization/travel insurance.

Further information

DVLA (2016, updated 2019) Assessing fitness to drive: a guide for medical professionals. 🖱 www.gov.uk/government/publications/assessing-fitness-to-drive-a-guide-for-medical-professionals

NICE (2015, updated 2016) Type 1 diabetes in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng17

NICE (2015, updated 2017) Type 2 diabetes in adults: management. 🖱 www.nice.org.uk/guidance/ng28

Patient advice and support

Diabetes UK ☎ 0345 123 2399 🖱 www.diabetes.org.uk

Treatment of type 2 diabetes

⚠ *Always* combine drug treatment of hyperglycaemia with lifestyle measures and modification of other risk factors for vascular complications.

Healthy eating and exercise Diet is the cornerstone of DM treatment. Diet sheets are available from Diabetes UK. ↑ physical activity is also beneficial (↓ weight, ↓ lipids, and ↑ insulin sensitivity).

Usage of antidiabetic drugs Figure 11.2

Antidiabetic drugs

Biguanides e.g. metformin 500mg tds. ↓ gluconeogenesis and ↑ peripheral utilization of glucose. Only effective if some endogenous insulin. Hypoglycaemia is not a problem. Start with minimum dose and ↑ every month until control is achieved/maximum dose reached. If not tolerated, try MR preparation. Avoid/stop if eGFR <30mL/min/1.73m^2.

Sulfonylureas (SU) e.g. gliclazide 80–160mg bd. Augment insulin secretion; only effective if some endogenous insulin production. All are equally effective. Advise to take before meals—warn about hypoglycaemia if meals are omitted and need for blood monitoring if driver. Start at minimum dose and ↑ until blood sugar is controlled/maximum dose is reached. Wait ≥1mo between adjustments. Main side effect is weight ↑.

Dipeptidylpeptidase-4 inhibitors (DPP4i) 'Gliptins', e.g. sitagliptin 100mg od, linagliptin 5mg od (useful if renal failure as excreted via the gallbladder). Inhibit breakdown of glucagon-like peptide-1 (GLP-1). This ↓ blood glucose levels by ↑ insulin secretion from the pancreas, ↓ glucagon secretion, and slowing gastric emptying. May cause hypoglycaemia (uncommon).

GLP-1 mimetics e.g. exenatide, liraglutide, lixisenatide. ↑ insulin secretion, ↓ glucagon secretion, and slow gastric emptying. Consider with metformin + sulfonylurea if triple therapy has failed and weight ↓ might benefit other co-morbidities, BMI ≥35kg/m^2 or BMI <35kg/m^2 but insulin treatment would have significant occupational implications. May cause hypoglycaemia—warn about the need for blood glucose monitoring if driver. Avoid if eGFR <30mL/min/1.73m^2 (<50 for some MR preparations). Continue after 6mo only if HbA1c has ↓ >11mmol/mol and weight ↓ is >3% of initial body weight. ❗ Rarely associated with ketoacidosis and pancreatitis.

Pioglitazone 15mg od. ↓ peripheral insulin resistance. Does not cause hypoglycaemia. May ↑ weight. Check LFTs before and regularly during treatment. ❗ ↑ risk of bladder cancer, bone fracture, fluid retention, and heart failure; avoid if risk factors for these conditions.

Sodium–glucose cotransporter-2 inhibitors (SGLT2i) 'Gliflozins', e.g. canagliflozin 100–300mg od; dapagliflozin 10mg od; empagliflozin 10–25mg od. Act on the proximal tubules of the kidney to block reabsorption of glucose back into the bloodstream resulting in ↓ blood glucose. Cause weight ↓; may cause UTIs and/or genital thrush. Avoid if aged >75y or

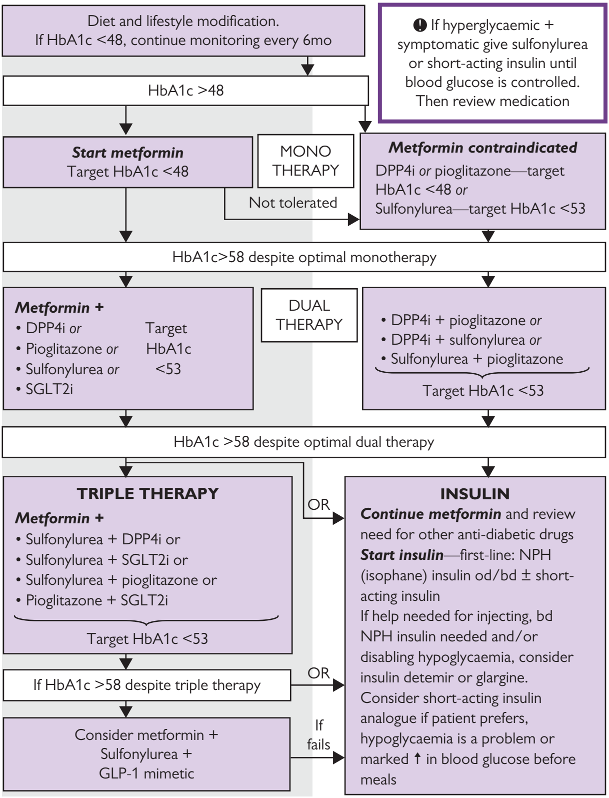

Diet and lifestyle modification.
If HbA1c <48, continue monitoring every 6mo
❗ If hyperglycaemic + symptomatic give sulfonylurea or short-acting insulin until blood glucose is controlled. Then review medication
HbA1c >48
Start metformin
Target HbA1c <48
MONO THERAPY
Not tolerated
Metformin contraindicated
DPP4i or pioglitazone—target HbA1c <48 or
Sulfonylurea—target HbA1c <53
HbA1c>58 despite optimal monotherapy
Metformin +
• DPP4i or
• Pioglitazone or
• Sulfonylurea or
• SGLT2i
Target HbA1c <53
DUAL THERAPY
• DPP4i + pioglitazone or
• DPP4i + sulfonylurea or
• Sulfonylurea + pioglitazone
Target HbA1c <53
HbA1c >58 despite optimal dual therapy
OR
TRIPLE THERAPY
Metformin +
• Sulfonylurea + DPP4i or
• Sulfonylurea + SGLT2i or
• Sulfonylurea + pioglitazone or
• Pioglitazone + SGLT2i
Target HbA1c <53
If HbA1c >58 despite triple therapy
OR
Consider metformin + Sulfonylurea + GLP-1 mimetic
If fails
INSULIN
Continue metformin and review need for other anti-diabetic drugs
Start insulin—first-line: NPH (isophane) insulin od/bd ± short-acting insulin
If help needed for injecting, bd NPH insulin needed and/or disabling hypoglycaemia, consider insulin detemir or glargine.
Consider short-acting insulin analogue if patient prefers, hypoglycaemia is a problem or marked ↑ in blood glucose before meals

Figure 11.2 Usage of antidiabetic drugs[N] (HbA1c units are in mmol/mol)

eGFR <60mL/min/1.73m^2. ❗ Rarely may cause diabetic ketoacidosis even with normal blood glucose—warn patients.

Rapid-acting insulin secretagogues e.g. repaglinide, nateglinide. Stimulate insulin release. Rapid onset and short duration of activity. Take ½ h before meals. Avoid if aged >75y. Hypoglycaemia is a common side effect. Warn about need for blood glucose monitoring if driver.

Starting insulin ➔ p. 322.

Lifestyle, drug and surgical treatment of obesity ➔ p. 152.

Further information

NICE (2015, updated 2017) Type 2 diabetes in adults: management. 🖱 www.nice.org.uk/guidance/ng28

Insulin

First-line treatment for type 1 DM and used when diet plus oral therapy have failed for type 2 DM. Local guidelines govern who does what.

⚠ All drivers must notify the DVLA and their insurance company.

Insulin passports and patient information booklets Offer to all patients receiving insulin. Provides a record of the patient's current insulin preparations and contains a section for emergency information. The patient information booklet provides advice on the safe use of insulin.

Types of insulin

- Rapid-acting analogues e.g. insulin lispro—fastest-acting; peak 0–3h after injection; last 2–5h; given just prior to meals
- Soluble (clear) human, porcine, or bovine e.g. Actrapid®—short-acting; peak 2–6h after injection; last 8h; give 15–30min before meals
- Isophane (NPH, cloudy) human, porcine, or bovine e.g. Humulin I®, Insulatard®. Intermediate- or long-acting. Peak action 4–12h after injection; lasts up to 30h. Taken od/bd to provide background insulin
- Long-acting insulin analogues e.g. insulin detemir or glargine—last 24h; provide background insulin; associated with ↓ risk of hypoglycaemia
- Premixed Combination of short- + long-acting insulin, e.g. 30%:70%

Injection regimens^N

Type 1 DM Offer a multiple daily injection basal-bolus regimen as first line. Use insulin detemir bd as basal insulin therapy (od insulin detemir or insulin glargine are alternatives) and a rapid-acting insulin analogue before meals. *Other options include*: bd human mixed insulin regimen if multiple daily injections are not possible (or bd analogue mixed insulin if a human mixed regimen causes unacceptable hypoglycaemic episodes).

❗ Consider adding metformin to insulin therapy for adults with type 1 DM and BMI ≥25kg/m^2 (or ≥23kg/m^2 if of South Asian origin).

Type 2 DM Continue to offer metformin (if already taking). Review continued need for other antidiabetic drugs. *Suitable insulin regimens*:

- Once/twice-daily intermediate/long-acting NPH insulin—consider adding short-acting human insulin separately or as pre-mixed preparation if HbA1c ≥75mmol/mol or HbA1c targets are not met
- Once-daily long-acting analogue (e.g. insulin detemir/glargine) is an alternative if help is needed to inject or if unwilling to inject bd
- If unacceptable hypoglycaemic episodes, consider an insulin analogue (e.g. insulin detemir/glargine) instead of human insulin. Biphasic preparations containing short-acting insulin analogues are also useful if blood glucose levels ↑ markedly after meals

Administration Deep sc injection into upper arm, thigh, buttock, or abdomen. Fat hypertrophy and scarring are minimized by rotation of injection sites. 'Pen' devices and syringe/needle are equally effective. Prime the needle using an 'air shot' (an empty needle ↓ insulin dose by ~2 units). Rock 'pens' containing pre-mixed insulins to mix contents. ↑ absorption can occur if a limb is exercised following injection.

Insulin pumps Continuous infusion of rapid-acting insulin may be considered in specialist settings if type 1 DM and ≥12y with repeated/unpredictable hypoglycaemia or inadequate glycaemic control (HbA1c >69mmol/mol) despite optimized multiple injection regimen, *or* <12y and multiple injection regimens are impractical or inappropriate.

Monitoring Check blood glucose preprandially ≥1×/d at different times—more often if multiple injection regimens, after dose changes, or during intercurrent illness; ask patients to keep a timed/dated diary of readings (many meters do this automatically). Record episodes of hypoglycaemia. *Target*: blood glucose 4–7mmol/L pre-meals with hypoglycaemic episodes kept to a minimum (4–8mmol/L pre-meals if <18y old).

Starting insulin for patients with type 2 DM Use a structured programme. Appropriate training is needed for all practice staff involved.

- Before starting—teach home blood glucose monitoring; reinforce diet
- Continue metformin ± other antidiabetic drugs
- Teach insulin injection technique—start 10 units of intermediate- or long-acting insulin od
- Teach patients/carers about safe disposal of sharps and hypoglycaemia
- Give instructions to ↑ dose every 3–7d until target levels are reached, e.g. average fasting glucose >10mmol/L—↑ by 6–8 units/d; 8–10mmol/L—↑ by 4–6 units/d; 6–8mmol/L—↑ by 2–4 units/d
- Provide a contact telephone number for advice; follow-up after 2–3d and then as needed; check HbA1c every 3mo until stable

Exercise ↓ insulin dose acting at the time of exercise *or* take 1–2 glucose tablets before exercise then check blood glucose afterwards. Adjust alterations/glucose dose with experience of effects of exercise.

Intercurrent illness Continue insulin in usual dose and keep a regular check (≥qds) of blood glucose. Consider blood/urine ketone monitoring for adults with type 1 DM. Maintain glucose intake even if not eating (with, e.g. Lucozade® or milk). If glucose >13mmol/L, ↑ insulin by 2 units/d until control is achieved or use top-up injections of short-acting insulin qds prn. *Admit to hospital if*: condition warrants admission; unable to take glucose; persistent vomiting and/or dehydration; ketotic (check urine if blood sugar >13mmol/L).

Poor control

- Exclude intercurrent illness
- Consider psychosocial factors
- Consider diet and/or gastroparesis
- Check insulin is being used correctly
- Check injection sites are not scarred or hypertrophic
- Consider changing insulin dose—ask the patient to record a glucose profile (blood sugar pre-meals and before bed)—if using >1 insulin adjust 1 at a time ↑ or ↓ as needed; alter by ≤10% each time; allow ≥48h between dose adjustments; alter dose of insulin acting at the time blood sugar is most out of control

Further information

NICE (2015, updated 2016) Type 1 diabetes in adults: diagnosis and management. 🖰 www.nice.org.uk/guidance/ng17

NICE (2015, updated 2017) Type 2 diabetes in adults: management. 🖰 www.nice.org.uk/guidance/ng28

Hypoglycaemia and diabetic renal disease

Hypoglycaemia Suspect if known diabetes on oral/insulin therapy. Short history. Warning signs/symptoms are usually present—sweating, hunger, tremor. If untreated, may progress to odd/violent behaviour, fits, and ultimately coma. On examination, the patient may be sweaty, and have tachycardia ± ↑ BP. ❗ Younger children may present atypically with behavioural changes or headache.

Emergency management Check capillary blood glucose on a blood testing strip. Suspect hypoglycaemia if <2.5mmol/L. If confirmed:

- If conscious give simple carbohydrate, e.g. 3 glucose tablets, 100mL of milk or a sugar-containing soft drink, e.g. Lucozade®, 5 sweets (e.g. Jelly Babies®), or GlucoGel®
- If unable to take oral carbohydrate, give IM glucagon 1mg (children <25kg—0.5mg). Takes ≤5min to act but may have poor effect if the patient is starved or drunk. Alternatively (if available) give IV glucose (adult: 50–250mL of 10% solution in 50mL aliquots; child: 2–5mL/kg of 10% solution)
- Once the patient has regained consciousness supplement with simple carbohydrate as for the conscious patient and, as symptoms improve, give complex carbohydrate, e.g. biscuits
- Repeat glucose testing in <15min then monitor hourly blood sugars over the next 4h and 4-hourly for the following 24h
- Maintain a high glucose intake for several hours if the patient has a severe episode of hypoglycaemia due to an oral medication
- Review reasons for the hypoglycaemia

Advice for patients

- Check blood sugar before driving and every 2h during a long journey
- Carry glucose everywhere and sandwiches on long journeys
- If warning signs of hypoglycaemia occur, stop hazardous activities and take action
- Wait until fully recovered (usually ~45min) before resuming activities

In case of severe hypoglycaemia Supply a responsible member of the family with glucose gel (e.g. GlucoGel®) and glucagon injection—teach him/her to use it. Response is short-lived—give oral glucose (e.g. Lucozade®, glucose tablets, milk) as soon as the patient is conscious.

Recurrent hypoglycaemia If hypoglycaemia occurs in a regular pattern, check pattern of meals and activity and alter insulin to match needs. If erratic, consider erratic lifestyle, alcohol, problems with absorption, errors in administration, and/or gastroparesis. If no obvious cause, consider change in underlying insulin sensitivity (e.g. age, CKD).

Hypoglycaemia unawareness To restore warning signs adjust insulin/food intake to stop glucose levels dropping to <4mmol/L. Consider undetected night-time hypoglycaemia if HbA1c is lower than expected from blood sugar diary.

❗ Driving is not permitted if hypoglycaemic awareness has been lost or >1 episode of severe hypoglycaemia while awake in the past 12mo (no episodes permitted for Group 2 licence).

Renal disease

Urinary tract infections More common in patients with poorly controlled DM. May exacerbate renal failure and → renal scarring. Consider papillary necrosis if recurrent (more common in DM).

Diabetic nephropathy Most common cause of end-stage renal failure in adults starting dialysis in the UK. 25% of people with DM have renal damage—more common if of Asian or African ethnic origin. Characterized by proteinuria, ↑ BP, and progressive ↓ in renal function. Classification of CKD (➔ p. 411).

Testing for nephropathy Before overt nephropathy occurs, there is a phase (microalbuminuria) in which the urine contains traces of protein not detected by standard protein dipstick. Presence of ↑ urine albumin levels and/or ↑ serum creatinine is associated with ↑ CVD risk and ↑ risk of CKD. Check estimated glomerular filtration rate (eGFR) and urinary albumin:creatinine ratio (ACR) at least annually if type 1 DM for >5y or type 2 DM of any duration. Check more frequently as needed if known CKD—➔ p. 414.

Management of nephropathy Discuss causes of CKD, risks (progression of renal disease, ↑ BP, CVD, ↑ mortality), and management:

- Optimize blood glucose control; stop/avoid nephrotoxic drugs (e.g. NSAIDs). Consider ↓ dose of other drugs as excretion/metabolism may be impaired. ❗ Stop metformin if eGFR <30 mL/min/1.73m^2
- Treat ↑ BP—target <130/80mmHg; offer ACE inhibitor/ARB if CKD + DM + ACR ≥3mg/mmol; offer atorvastatin 20mg nocte for primary prevention to all patients with DM + CKD (↓ CVD events and death by 20%). ↑ dose if <40% ↓ in non-HDL cholesterol and eGFR ≥30mL/min/1.73m^2. If eGFR <30mL/min/1.73m^2, seek specialist advice
- Offer folic acid/vitamin B_{12} supplements if ↓ on laboratory testing/poor diet

Refer for renal USS If:

- eGFR of <30 mL/min/1.73m^2 (G4/5) or accelerated progression of CKD: sustained ↓ in eGFR of ≥25% and a change in GFR category in <12mo, or sustained ↓ in eGFR of ≥15mL/min/1.73m^2 per year
- Symptoms of urinary tract obstruction
- Family history of polycystic kidney disease and aged >20y

Refer to renal physician If:

- eGFR <30mL/min/1.73m^2 *or* ACR ≥70mg/mmol *or* ACR ≥30mg/mmol + haematuria (unless urgent referral for suspected cancer is indicated—➔ p. 420)
- Sustained ↓ in eGFR of ≥25% + change in GFR category or sustained ↓ in eGFR of ≥15 mL/min/1.73m^2 in <12mo
- Poorly controlled BP despite ≥4 antihypertensive drugs
- Known/suspected rare/genetic causes of CKD or renal artery stenosis

Further information

NICE (2014, updated 2015) Chronic kidney disease in adults. 🖱 www.nice.org.uk/guidance/cg182

NICE (2015, updated 2016) Type 1 diabetes in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng17

Diabetic complications: cardiovascular and skin

Cardiovascular complications Diabetic patients are at ↑ risk of MI (2–5×), stroke (2–3×), and peripheral vascular disease. Protective effect of female sex is lost. Atherosclerotic disease accounts for most of the excess mortality due to DM. Check arterial risk factors annually:

- Age
- Family history of arterial disease
- Abdominal adiposity
- BP
- Lipid profile (LDL, HDL cholesterol, and triglycerides)
- Albumin excretion rate
- Blood glucose control
- Smoking—give smoking cessation advice at every opportunity. Help patients who want to give up with advice, medication, and support

Blood glucose ➲ p. 317. Target HbA1c:

- <48mmol/mol if type 1 DM or type 2 DM controlled with diet and lifestyle alone ± a single drug not associated with hypoglycaemia, e.g. metformin, a DPP4i, or pioglitazone
- <53mmol/mol if type 2 DM treated with a single drug associated with hypoglycaemia or any combination of antidiabetic drugs

Statin ➲ p. 223. Consider statin therapy for:

Primary prevention If:

- >85y (if appropriate)
- 10y CVD risk ≥10%—use a risk calculator that includes type 2 DM in its risk calculation, e.g. QRisk3
- Type 1 DM + >40y or DM for >10y or established nephropathy or other CVD risk factors
- eGFR <60 mL/min/1.73 m^2 and/or albuminuria

Start treatment after optimizing lifestyle intervention, and treatment of other modifiable risk factors/secondary causes of dyslipidaemia. Start with high-intensity statin (e.g. atorvastatin 20–40mg od). ↑ dose as needed aiming for a 40% ↓ non-HDL cholesterol from baseline.

Secondary prevention If history of CVD—start treatment immediately irrespective of initial cholesterol levels. Use a high intensity statin (e.g. atorvastatin 80mg od).

BP ➲ p. 218. Any ↓ in average BP ↓ risk of cardiovascular complications. Measure BP annually if not hypertensive and no renal disease. If BP is higher than target, consider 24h BP monitoring.

- **Type 1 DM**[N] Target BP is ≤135/85mmHg unless microalbuminuria/proteinuria or ≥2 features of the metabolic syndrome (➲ p. 313) when treated if BP >130/80mmHg
- **Type 2 DM**[N] Target BP is <140/80mmHg or <130/80mmHg if kidney, eye or cerebrovascular disease

Choice of antihypertensive[N]

- In all cases, discuss lifestyle modifications (➲ p. 219)
- If ↑ BP, start with an ACE inhibitor. If side effects with ACE inhibitor, ARB is an alternative. For people of African-Caribbean descent, offer ACE inhibitor + Ca^{2+} channel blocker or diuretic. If possibility of

becoming pregnant, start with a Ca^{2+} channel blocker. Monitor BP every 1–2mo until stable within target. Titrate dose to maximum tolerated
- If BP remains above target add a Ca^{2+} channel blocker (e.g. amlodipine 5mg od) and/or a diuretic (e.g. indapamide MR 1.5mg od)
- Fourth line agents include α-blockers, β-blockers, or further diuretic therapy, e.g. spironolactone 25mg od. Consider referral

Monitoring Monitor BP every 4–6mo once stable on treatment. Check for drug side effects (including erectile dysfunction and postural drop).

Antiplatelet therapy Give aspirin or clopidogrel 75mg od to all those with a prior history of CVD. Do not use for 1° prevention.

Skin changes associated with DM Include:
- Predisposition to infection, e.g. candidiasis, staphylococcal infection
- Pruritus
- Xanthomas
- Diabetic bullae
- Vitiligo (type 1 DM)
- Fat atrophy/hypertrophy at insulin injection sites
- Acanthosis nigricans
- Neuropathic and/or ischaemic ulcers—➔ p. 330
- Psoriasis—people with psoriasis have a 21% ↑ risk of type 2 DM
- Necrobiosis lipoidica (Figure 11.3a)—50% associated with DM. Small, dusky red, well-circumscribed nodule(s), usually on the shin. Enlarge slowly becoming brownish yellow, irregular, and flattened/depressed. Long-standing lesions may ulcerate. No effective treatment
- Diabetic dermopathy (Figure 11.3b)—pigmented scars over shins
- Diabetic cheiroarthropathy—waxy skin-thickening over the dorsum of the hand with restricted mobility
- Granuloma annulare 💣—asymptomatic dermal nodules—association with DM is controversial

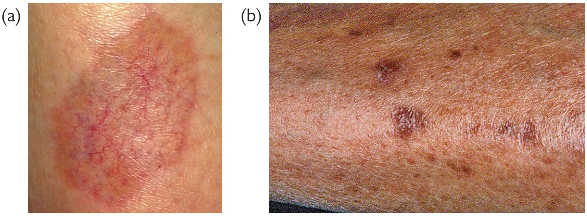


Figure 11.3 Diabetic skin changes: (a) necrobiosis lipoidica and (b) diabetic dermopathy

Figure 11.3 (a) reproduced with permission from New Zealand Dermatological Society Incorporated, courtesy of Prof Raimo Suhonen. Published online at: www.dermnetnz.org

Further information

NICE (2014, updated 2016) Cardiovascular risk assessment and the modification of blood lipids for the primary and secondary prevention of cardiovascular disease. 🖱 www.nice.org.uk/guidance/cg181

NICE (2011, updated 2016) Hypertension in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg127

NICE (2015, updated 2016) Type 1 diabetes in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng17

NICE (2015, updated 2017) Type 2 diabetes in adults: management. 🖱 www.nice.org.uk/guidance/ng28

Diabetic complications: eye and nerve

Eye disease ~1 in 3 people with diabetes have eye problems at the time of diagnosis.

Blurred vision May occur if control is poor—caused by osmotic changes in the lens, corrects with normalization of blood sugar. Wait before changing glasses.

Cataract Juvenile 'snowflake' cataracts are more common and can develop rapidly (over days). Senile cataracts occur ~10y earlier in DM. Surgical complications from cataract removal are more common due to corneal epitheliopathy related to DM.

Retinopathy Most common cause of blindness in people of working age in industrialized countries (risk is ↑ ×20 compared to non-diabetics). 20–40% patients with type 2 DM have retinopathy at diagnosis. 20y after diagnosis, 95% with type 1 DM, and 60% with type 2 DM have retinopathy—sight-threatening in 5–10%.

Pathogenesis of retinopathy Small retinal blood vessels become blocked, swollen (aneurysms), or leaky causing exudate formation, oedema, or new vessels. Often asymptomatic until late stages. Laser treatment (photocoagulation) halts progression but does not restore vision. Good diabetic control slows development of retinopathy. Monitor and treat risk factors—BP, lipids (hard exudates), smoking.

Classification of retinopathy Various classifications predict prognosis. All are based on whether new vessels are present/absent and if the macula is affected:
- **Non-proliferative or background retinopathy** No new vessels; further graded by severity (number of microaneurysms)
- **Proliferative retinopathy** New vessels present; further classified by location of new vessels (how close to the optic disc) and severity
- **Maculopathy** Involvement of the macula

> ❗ *Retinal screening* Digital retinal photography is available throughout the UK. Ensure patients are referred early for retinal screening (<3mo after diagnosis) and are screened ≥1×/y to detect retinopathy before visual loss occurs. Screening in pregnancy—➜ p. 806.

Glaucoma and rubeosis iridis DM is not a risk factor for primary glaucoma but glaucoma is more likely to be found in patients with DM due to regular eye checks. Rubeosis iridis is the growth of new vessels on the iris in eyes with advanced retinal ischaemia. This predisposes to a severe form of secondary angle-closure glaucoma.

Retinal detachment More common in patients with proliferative diabetic retinopathy. Caused by contraction of the vitreous gel in association with haemorrhage from new vessels and subsequent fibrosis.

Other eye conditions more common in DM Optic neuropathy (type 2 DM; due to vascular occlusion); retinal vein occlusion; ocular nerve palsies.

Refer to ophthalmology if E = Emergency; *U* = Urgent; *R* = Routine

- Sudden loss of vision—*E*
- Rubeosis iridis—*E*
- Pre-retinal or vitreous haemorrhage—*E*
- Retinal detachment—*E*
- New vessel formation—*U*
- Maculopathy—*R*
- Pre-proliferative retinopathy—*R*
- Cataract affecting visual acuity—*R*
- Unexplained drop in visual acuity—*R*

Neuropathy Enquire annually about painful and other symptomatic neuropathy, erectile dysfunction in men, and manifestations of autonomic neuropathy especially if renal complications or erratic blood glucose control. Optimize blood glucose control.

Symmetrical sensory progressive polyneuropathy Affects 40–50% of patients with DM eventually. Starts distally feet > hands. Glove-and-stocking distribution. May be asymptomatic or cause numbness, tingling, or neuropathic pain. Pain can be depressing and disabling. Be supportive. If simple analgesia with paracetamol or NSAID is ineffective, try neuropathic painkillers (➔ p. 188). When pain is controlled, review regularly and consider reducing dose/stopping.

Mononeuropathies/mononeuritis multiplex Especially cranial nerves III and VI resulting in ocular palsies—➔ p. 512

Amyotrophy Painful wasting of quadriceps muscles—reversible with improved blood sugar control.

Autonomic neuropathy

- **Postural ↓ BP** Fall of >20mmHg systolic (or >10mmHg diastolic) BP on standing. Common especially in the elderly. Review medication and stop (if possible) drugs that may be contributing. ↑ dietary salt intake may help. Other treatments include fludrocortisone 100–400 mcg od (unlicensed—uncomfortable oedema is a common side effect), and midodrine (2.5–10mg tds)
- **Gastric paresis** Treat with an antiemetic which promotes gastric transit, e.g. domperidone 30mg tds. 💣 Use of erythromycin for treatment of gastroparesis is controversial[N]
- **Diabetic diarrhoea** Common. Exclude other causes of change in bowel habit—➔ p. 378. Otherwise treat with loperamide 2mg prn
- **Gustatory sweating** Can be treated with antimuscarinics (e.g. propantheline) but side effects are common. Hyperhidrosis—➔ p. 575
- **Urinary retention** ➔ p. 428
- **Erectile dysfunction** ➔ p. 754

Depression Prevalence of depression is ↑ in patients with DM. Screen for depression as part of the annual diabetic check (➔ p. 173)

Further information

NICE (2013) Gastroparesis in adults: oral erythromycin. 🖱 www.nice.org.uk/advice/esuom13/chapter/Key-points-from-the-evidence
NICE (2015, updated 2016) Type 1 diabetes in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng17
NICE (2015, updated 2017) Type 2 diabetes in adults: management. 🖱 www.nice.org.uk/guidance/ng28

The diabetic foot

Foot problems are common in diabetes. Diabetic foot ulcers precede >80% of amputations and DM is the most common cause of non-traumatic limb amputation. 5-year mortality is ~70% after an amputation. Foot problems result from:

- Peripheral neuropathy → ↓ foot sensation (Table 11.3) *and*
- Peripheral vascular disease → pain and predisposition to ulceration (Table 11.3)

❗ Patients with diabetes may have coexisting peripheral neuropathy and peripheral vascular disease.

Information about foot care

- Self-care and self-monitoring:
  - Daily examination of the feet for problems—colour change; swelling; breaks in the skin; numbness
  - Footwear—importance of well-fitting shoes and hosiery
  - Hygiene (daily washing and careful drying) and nail care
  - Dangers associated with procedures, e.g. corn/verruca removal
  - Wound care
  - Methods to help self-monitoring, e.g. mirrors if ↓ mobility
- When to seek advice from a health professional—if any colour change, swelling, breaks in the skin or numbness, or if self-monitoring is not possible (e.g. due to mobility problems)
- For patients at moderate/high risk or with ulcers, additionally advise no barefoot walking and that, due to ↓ sensation, extra care and attention is needed, particularly with footwear
- If skin lesions, advise patients to seek help if any change in the lesion, if ↑ swelling, pain, odour, colour change or systemic symptoms

Risk factors

- Age >70y
- Plantar callus
- Neuropathy
- Long DM duration
- Peripheral vascular disease
- Previous ulceration or amputation
- Social deprivation/isolation
- Foot deformity
- Poor footwear
- Poor vision
- Smoking

The diabetic foot check Part of the annual diabetic review.

History

- Foot problems since last review
- Visual or mobility problems affecting self-care of feet
- Self-care behaviours and knowledge of foot care
- History of numbness, tingling, or burning—may be worse at night

Table 11.3 Clinical features of neuropathic and vascular foot ulcers

Neuropathic	Vascular
Warm foot	Cool foot
Bounding pulses, normal ABPI	Absent pulses, ↓ ABPI (❗ may be normal or ↑ due to calcification of vessels)
Located at pressure points	Located at extremities (e.g. between toes)
Painless	Painful
Clearly defined or 'punched out'	Less clearly delineated
Surrounded by callus	

Table 11.4 Classification of foot risk

Foot risk	Features
Low risk	No risk factors except callus alone
Moderate risk	Deformity *or* neuropathy *or* non-critical limb ischaemia
High risk	• Previous ulceration or amputation *or* • On renal replacement therapy *or* • Neuropathy and non-critical limb ischaemia together *or* • Neuropathy in combination with callus and/or deformity *or* • Non-critical limb ischaemia in combination with callus and/or deformity
Active diabetic foot problem	• *Skin*: ulceration or spreading infection *or* • *Vascular*: critical limb ischaemia or gangrene *or* • *Joint*: suspicion of an acute Charcot arthropathy, or an unexplained hot, red, swollen foot ± pain

Examination

- Foot shape, deformity, joint rigidity, and shoes
- Skin condition—fragility, cracking, oedema, callus, ulceration, sweating, presence of hair
- Foot and ankle pulses ± ABPI
- Sensitivity to 10g monofilament

Management

General points Optimize diabetic control and risk factors for vascular disease (including smoking cessation); review drug therapy—stop β-blockers if peripheral vascular disease; educate about foot care.

Specific management Classification—Table 11.4.

- Low risk Annual foot assessments. Education about foot care and risk of progression to moderate/high risk
- Moderate/high risk Refer to the foot protection service to be seen in <4wk if high risk and <8wk if moderate risk. Reassess feet every 3–6mo if moderate risk; every 1–2mo if high risk or more frequently if concern
- Active diabetic foot problem If limb/life-threatening condition (Box 11.1), admit as a same-day emergency; otherwise, refer for assessment in <1 working day to the multidisciplinary foot care service

⚠ Box 11.1 Limb-or life-threatening foot changes requiring immediate hospital admission

- Ulceration + fever/sepsis
- Ulceration + limb ischaemia
- Clinical concern about deep soft tissue infection/osteomyelitis
- Gangrene

Charcot osteoarthropathy (Charcot's joint) Neuropathic foot damaged because of trauma 2° to loss of pain sensation. Manage as for 'Active diabetic foot problem'.

Further information

NICE (2015, updated 2016) Diabetic foot problems: prevention and management. 🖱 www.nice.org.uk/guidance/ng19

Lumps in the thyroid gland and goitres

Faced with a lump in the pre-tracheal region of the neck, ask:

- Is it in the thyroid (moves up and down on swallowing)?
- Is it a solitary lump or more generalized (a goitre)?
- Is the patient thyrotoxic, euthyroid, or hypothyroid?
- Is the trachea being compressed (patient is breathless or stridor)?

⚠ Urgent management of thyroid lumps

Refer to be seen immediately by a thyroid surgeon If symptoms of tracheal compression including stridor due to thyroid swelling.

Refer urgently to a thyroid surgeon[N] (to be seen in <2wk) If any age and unexplained thyroid lump.

Solitary thyroid nodule Investigate *all* solitary nodules. Check TFTs and refer urgently to exclude thyroid cancer. Differential diagnosis:

- Benign (~90%): cyst, adenoma, discrete nodule in a nodular goitre
- Malignant (~10%): *primary*—thyroid adenocarcinoma, lymphoma, medullary carcinoma; *secondary*—direct spread from local tumour, metastatic spread from breast, colon/rectum, kidney, lung, lymphoma

Carcinoma of the thyroid Primary tumours:

- Papillary adenocarcinoma (60%) Typical age: 10–40y. ♀ > ♂. Low-grade malignancy. Rarely fatal. Spreads to local LNs and/or lung. Sensitive to TSH. Treated with thyroidectomy then lifelong thyroxine
- Follicular carcinoma (25%) Typical age range: 40–60y. ♀ > ♂. May arise in a pre-existing multinodular goitre. Spreads via bloodstream. Bony secondaries are common. Treatment is with surgery and thyroxine suppression therapy and/or radioactive iodine
- Lymphoma (5%) Occurs at any age. Involvement of the thyroid may be 1° or 2°. Associated with Hashimoto's thyroiditis. Staged/treated as for lymphoma elsewhere (➔ p. 656). Prognosis is good
- Anaplastic carcinoma (rare) Typical age: 50–60y. ♀ > ♂. Aggressive tumour. Grows rapidly and infiltrates tissues of the neck. Tracheal compression is common. Metastasizes locally to LNs and via lymphatics. Poor response to treatment
- Medullary carcinoma (rare) Occurs at any age. ♀ = ♂. Familial incidence; associated with adenomas elsewhere. Often secretes calcitonin (used as tumour marker). Spreads to local LNs. Treated by excision then chemotherapy ± radiotherapy

Thyroid adenoma Benign tumours of the thyroid. 4 types classified according to histological appearance—papillary, follicular, embryonal, Hürthle cell. A few produce thyroxine → thyrotoxicosis. Haemorrhage is rare and results in rapid ↑ in size. Refer for confirmation of diagnosis ± surgery.

Goitre There are 4 main types of goitre—Table 11.5.

Thyroid cyst Usually degenerative part of a nodular goitre though true cysts do occur. Rapid enlargement/pain may be caused by haemorrhage into a cyst. Refer for confirmation of diagnosis.

Table 11.5 Types of goitre—presentation and management

Type	Features	Management
Congenital	Enlarged thyroid gland present at birth ± hypo- or hyperthyroidism	Hypothyroid babies are treated with thyroxine; if there is tracheal compression or hyperthyroidism, treatment is surgical
Physiological	Occurs at puberty, during pregnancy, and in conditions of iodine deficiency	Usually requires no treatment. If iodine deficient, treat with iodine supplements
Nodular	Benign enlargement of the thyroid gland with areas of hyperplasia and involution	No treatment is necessary unless: • Thyrotoxic • Compression of the neck structures → dyspnoea or dysphagia • Worried by cosmetic appearance • Focal ↑ in size or recurrent laryngeal nerve palsy (hoarseness)—suggests malignant change—refer for urgent review (in <2wk) If treatment is needed, refer to surgery or endocrinology depending on symptoms
Toxic	*Grave's disease*: smooth thyroid enlargement + thyrotoxicosis	See management of hyperthyroidism—➔ p. 334
Inflammatory	*Hashimoto's thyroiditis*: ♀ > ♂. Antibodies to thyroid tissue are produced. Initially goitre and thyrotoxicosis. Later myxoedema	In all cases refer to endocrinology for confirmation of diagnosis and management guidance
	De Quervain's thyroiditis: inflammation due to viral infection—usually Coxsackie virus. Acutely swollen, tender thyroid gland and transient thyrotoxicosis often preceded by sore throat/malaise. Settles spontaneously	
	Riedel's thyroiditis: rare. Thyroid becomes infiltrated by scar tissue → hypothyroidism ± recurrent laryngeal nerve palsy ± stridor	

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Thyroid disease

Interpretation of thyroid function test results Table 11.6

Table 11.6 Interpretation of thyroid function test results

Results of TFTs	Interpretation	Notes
TSH ↓, T_4↑	Hyperthyroid (thyrotoxic)	Occasionally T_4 is normal but T_3 ↑; request T_3 levels if low TSH and normal T_4
TSH ↑, T_4↓	Hypothyroid	TSH ↓ if hypothyroidism is secondary to pituitary failure (rare)
TSH ↑, T_4 normal	Subclinical hypothyroidism	If any symptoms (including depression and non-specific symptoms or hypercholesterolaemia) consider a trial of treatment. If no symptoms, repeat after 3–6mo and then monitor annually

Hyperthyroidism Affects 2% ♀ and 0.2% ♂. *Peak age*: 20–49y. *Causes*:

- Graves' disease
- Toxic nodular goitre—older ♀ with past history of goitre
- Thyroiditis
- Amiodarone
- Kelp ingestion

Presentation

- Weight loss
- Tremor
- Palpitations
- Hyperactivity
- AF
- Hyperhidrosis
- Eye changes
- Infertility
- Alopecia

❗ In elderly patients, symptoms may be less obvious and include confusion, dementia, apathy, and depression.

Management Refer to endocrinology at presentation. *Treatment*:

- **β-blockers** e.g. propranolol, atenolol. Useful for symptom control until antithyroid drug therapy takes effect
- **Carbimazole** Inhibits synthesis of thyroid hormones. Ineffective for treatment of thyroiditis. May be given short term to render a patient euthyroid prior to surgery or treatment with radioactive iodine, or long term (12–18mo) to induce remission (but >50% relapse). 3/1000 patients have serious adverse effects—agranulocytosis, hepatitis, aplastic anaemia, or lupus-like syndromes
- **Radioactive iodine (^{131}I)** Effects take 3–4mo to become apparent. Withdraw carbimazole >4d prior to treatment and do not restart until >3d after. Advise women of child-bearing age to avoid pregnancy for 4mo. Most become hypothyroid (sometimes years) after treatment. Monitor TFTs long-term. Associated with small ↑ risk of thyroid malignancy
- **Surgery** Partial or total thyroidectomy—reserved for patients with large goitres or who decline radioactive iodine. Carries risk of damage to recurrent laryngeal nerve or parathyroids

⚠ Warn *all* patients starting carbimazole to stop the drug and seek urgent medical attention if they develop sore throat or other infection.

Hyperthyroid crisis (thyrotoxic storm) ➔ p. 1083

Graves' disease Most common cause of hyperthyroidism. ♀:♂ ≈5:1. *Peak age*: 30–50y. Associated with smoking and stressful life events. Autoimmune disease in which antibodies to the TSH receptor are produced.

Clinical features Hyperthyroidism; diffuse goitre ± thyroid bruit due to ↑ vascularity; extra-thyroid features: thyroid eye disease—25–50% (bilateral in >90%); pretibial myxoedema—5%; thyroid acropachy (clubbing, finger swelling)—rare; onycholysis—rare.

Management As for hyperthyroidism.

Thyroid eye disease Presents with:

- Eye discomfort ± protrusion (exophthalmos and proptosis)
- Lid lag
- Double vision/ophthalmoplegia (especially of upward gaze)
- TFTs can be ↑ or normal.

Management Treat any hyperthyroidism. Refer to ophthalmologist. If ↓ acuity or loss of colour vision—refer urgently as there may be optic nerve compression.

Hypothyroidism (myxoedema) Common—10% ♀ >60y, ♀:♂ ≈8:1.

Causes Chronic autoimmune thyroiditis, post ^{131}I, thyroidectomy.

Presentation Onset tends to be insidious and may go undiagnosed for years. *Always* consider hypothyroidism when a patient has non-specific symptoms, depression, fatigue, lethargy, or general malaise. Other symptoms—weight ↑, constipation, hoarse voice, or dry skin/hair. Signs are often absent—there may be a goitre, slow-relaxing reflexes, or non-pitting oedema of the hands, feet, or eyelids.

Screening Check TFTs in patients:

- With persistent symptoms of tiredness/lethargy without clear cause
- On amiodarone or with a history of ^{131}I administration
- With hypercholesterolaemia, infertility, Turner's syndrome, depression, dementia, obesity, DM, or other autoimmune disease

Management Patients taking thyroxine replacement are entitled to apply for free prescriptions in England (➔ p. 113).

- <65y and healthy 50–100 mcg od levothyroxine. Re-check TFTs after 4–6wk. Adjust dose to keep TSH in the normal range. Once dose is stable and TSH is within normal range monitor annually and if symptomatic or worries about compliance
- If elderly or pre-existing heart disease Start 25mcg od levothyroxine and ↑ dose every 4–6wk according to TFTs. Consider adding propranolol if history of CHD as levothyroxine can provoke angina

Withdrawal of levothyroxine Usually needed lifelong. If diagnosis is in doubt stop and re-measure TFTs after 4–6wk.

Hypothyroid (myxoedema) coma ➔ p. 1083

Information for patients

British Thyroid Foundation 🖱 www.btf-thyroid.org

Hyper- and hypocalcaemia

⚠ **Checking Ca^{2+}** Take an *uncuffed* sample (to avoid falsely high readings) and correct for serum albumin—for every mmol/L less than 40, a correction of 0.02mmol/L should be added. For example:

Calcium 2.40 Corrected calcium $= (40 - 24) \times 0.02 + 2.4$
Albumin 24 $= 0.32 + 2.4 =$ **2.72**

Hypocalcaemia ↓ serum calcium (<2.15mmol/L). *Causes*:

- If phosphate ↑ CKD, hypoparathyroidism (may be congenital or 2° to thyroid or parathyroid surgery, or malignant infiltration), pseudohypoparathyroidism (insensitivity to parathyroid hormone)
- If phosphate normal or ↓ Vitamin D deficiency (osteomalacia, rickets), malabsorption, lack of calcium in diet, overhydration, pancreatitis

Presentation May be subtle. Includes:

- Tetany
- Irritability, depression or psychosis
- Perioral paraesthesia
- Carpo-pedal spasm (wrist flexion and fingers drawn together)
- Neuromuscular excitability (tapping over parotid causes facial muscles to contract—Chvostek's sign)

❗ Apparent hypocalcaemia may be an artefact of hypoalbuminaemia.

Management Check vitamin D levels. Supplement with calcium. Referral may be needed to investigate/treat the underlying cause.

Hypercalcaemia ↑ level of serum calcium (>2.55mmol/L). *Prevalence* ≈1 in 500; ♂: ♀ ≈1:3. Rare <age 50y.

Common causes (90%)

- Primary hyperparathyroidism
- Malignancy (10% tumours—usually myeloma, breast, lung, kidney, thyroid, prostate, ovary or colon)

Uncommon causes

- Chronic renal failure
- Familial benign hypercalcaemia
- Sarcoidosis
- Thyrotoxicosis
- Milk alkali syndrome
- Vitamin D treatment

Presentation Often very non-specific. May be an incidental finding. Other symptoms: '*bones, stones, groans, and abdominal moans*'.

- Tiredness
- Lethargy
- Weakness
- Mild aches and pains
- Anorexia
- Weight loss
- Low mood
- Stone formation
- Nausea/vomiting (often intractable)
- Polyuria and polydipsia
- Abdominal pain
- Constipation
- Confusion
- Corneal calcification

Management

- Treat according to cause (Figure 11.4)—malignancy (➔ p. 1014); hyperparathyroidism (➔ p. 337)
- If diagnosis is unclear, refer to endocrinology. Urgency depends on serum Ca^{2+} and severity of symptoms

⚠ Hypercalcaemia can be fatal. If Ca^{2+} >3.5mmol/L or severe symptoms, admit for lowering of Ca^{2+} with forced diuresis + IV bisphosphonate.

Hyperparathyroidism ↑ secretion of parathyroid hormone (PTH).

- **Primary hyperparathyroidism** Incidence 0.5/1000. Peak age 40–60y. ♀:♂ ≈2:1. Circulating level of PTH is inappropriately high. Most patients are hypercalcaemic (but may be normocalcaemic if coexistent vitamin D deficiency). Due to ↑ secretion of PTH from one or both parathyroid glands. Refer. Treatment is usually surgical. Drug treatment (e.g. with cinacalcet) may be an option if unsuitable for surgery
- **Secondary hyperparathyroidism** ↑ PTH in response to chronic hypocalcaemia or hyperphosphataemia. Treat the underlying cause
- **Tertiary hyperparathyroidism** Inappropriately ↑ PTH → ↑Ca^{2+}. Follows prolonged secondary hyperparathyroidism. Most common in patients with chronic kidney disease (especially if on dialysis) or chronic malabsorption. Treatment may be either surgical or medical (e.g. with cinacalcet or paricalcitol)

Familial benign hypercalcaemia Asymptomatic. Inherited condition in which serum calcium concentrations are mildly ↑ throughout life. Confirm (if possible) by demonstrating ↑ Ca^{2+} in other family members. No adverse consequences and no treatment needed.

Milk alkali syndrome Usually due to ingestion of OTC indigestion remedies (e.g. Rennie® tablets). Ca^{2+} levels revert to normal on stopping. Investigate the reason why the patient is taking these remedies (? peptic ulcer). Sometimes also caused by calcium supplements taken with bisphosphonates for prophylaxis of osteoporosis—stop the calcium supplement.

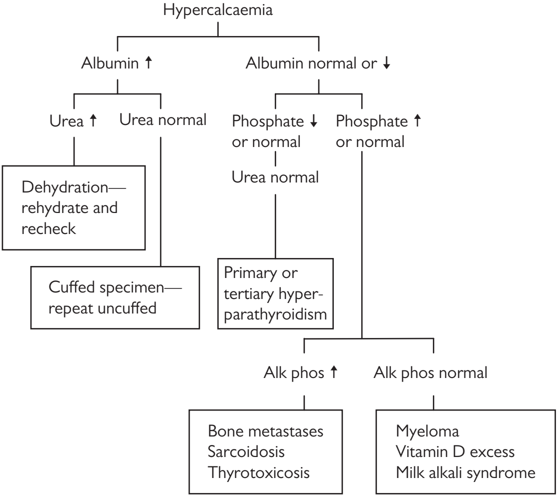

Hypercalcaemia
Albumin ↑
Albumin normal or ↓
Urea ↑
Urea normal
Phosphate ↓ or normal
Phosphate ↑ or normal
Dehydration—rehydrate and recheck
Urea normal
Cuffed specimen—repeat uncuffed
Primary or tertiary hyperparathyroidism
Alk phos ↑
Alk phos normal
Bone metastases
Sarcoidosis
Thyrotoxicosis
Myeloma
Vitamin D excess
Milk alkali syndrome

Figure 11.4 Guide to the diagnosis of cause of hypercalcaemia (must be taken in clinical context)

Adrenal disorders

Disorders of the adrenal cortex The adrenal cortex produces 3 classes of steroids:

- Glucocorticoids e.g. cortisol
- Mineralocorticoids e.g. aldosterone
- Sex hormones e.g. androstenedione, testosterone, and oestrogen

Disorders result from disturbance in production of these steroids:

Cushing's syndrome In the majority of cases, Cushing's syndrome is iatrogenic—caused by exogenous administration of prednisolone or other corticosteroids. Non-iatrogenic Cushing's syndrome is much rarer with annual incidence of 1–2/million (♀:♂ ≈3:1):

- 80% have a pituitary adenoma which secretes adrenocorticotrophic hormone (ACTH) causing hypersecretion of glucocorticoids and sex hormones (Cushing's disease)
- 20% are due to ectopic ACTH secretion by other tumours (e.g. small cell lung cancer) or hypersecreting tumours of the adrenal cortex

Presentation Cushing's syndrome has high morbidity and mortality. Clinical features include:

- Moon face (90%)
- Truncal obesity (85%)
- Hypertension (80%)
- Menstrual disturbance (80%)
- Striae and bruising (60%)
- Osteoporosis (60%)
- Lethargy/depression (60%)
- Hirsutism
- Acne
- Pigmentation
- DM
- Feminization in ♂
- Polyuria and polydipsia
- Psychosis

Management

- Stop/minimize exogenous steroids
- If no exogenous steroids and Cushing's syndrome is suspected, request a dexamethasone suppression test—dexamethasone 1mg po at midnight then serum cortisol measured at 9 a.m. If <50mmol/L excludes diagnosis unless cortisol secretion is episodic. If ≥50mmol/L, check ACTH level and 24h urinary free cortisol and seek expert advice

Adrenal insufficiency (Addison's disease) May be:

- Primary—resulting from adrenal disease/failure *or*
- Secondary—resulting from inadequate pituitary or hypothalamic stimulation of the adrenal glands

In the UK, most cases result from autoimmune disease, surgery, cessation of therapeutic corticosteroids, or failure to ↑ steroid dose to cover stress. Worldwide TB and AIDS are major causes.

Clinical features

- Tiredness (95%)
- Weakness (95%)
- Anorexia (95%)
- Weight loss (90%)
- Abdominal pain (30%)
- Myalgia/arthralgia (20%)
- Postural hypotension/fainting (15%)
- Nausea
- Pigmentation (buccal, palmar creases, new scars—90%)

Presentation Can be dramatic with coma and severe hypoglycaemia (➔ p. 1083—admit as an emergency) or insidious. 50% patients with autoimmune Addison's disease have or will develop another autoimmune disease (e.g. Graves' disease, pernicious anaemia) and 5% of women develop premature ovarian failure.

Short Synacthen® test Take 9 a.m. blood for serum cortisol levels; inject 250mcg Synacthen® (synthetic ACTH) IV or IM; take a further blood sample for serum cortisol levels ½h later. If 30min cortisol level is:

- >600nmol/L—adrenal insufficiency is excluded
- 400–590nmol/L—the result is equivocal—repeat
- <400nmol/L—adrenal insufficiency is confirmed—check ACTH (if ↓ investigate pituitary function; if ↑ investigate cause of adrenal disease)

Other investigations

- Biochemical abnormalities—↑ K^{+}, ↓ Na^{+}, ↓ glucose (may not be symptomatic), uraemia, ↑ Ca^{2+}, abnormal LFTs
- FBC—normocytic anaemia, eosinophilia, lymphocytosis

Management Refer to endocrinology, urgency depends on clinical state. Treatment usually involves replacing deficient steroids with hydrocortisone and fludrocortisone. ❗ Warn patients not to stop steroids abruptly, to tell any doctor treating them about their condition and wear Medic-Alert/Medi-Tag bracelet in case of emergency. Double dose of hydrocortisone prior to dental treatment or if intercurrent illness (e.g. URTI). If vomiting, replace hydrocortisone po with IM hydrocortisone.

Hyperaldosteronism Suggested by presence of ↑ BP resistant to treatment together with ↓ K^{+}—but normokalaemic cases are also described. May be primary (2 out of 3 have an aldosterone-secreting adenoma), when termed Conn's syndrome, or secondary to excess renin secretion (e.g. due to renal artery stenosis). If suspected, refer for endocrine assessment. Treatment depends on the cause. ❗ Rarely excess liquorice consumption can mimic Conn's syndrome.

Congenital adrenal hyperplasia ➔ p. 873

Disorders of the adrenal medulla

Phaeochromocytoma Rare but serious disorder affecting 0.1% hypertensive patients. Usually caused by catecholamine-secreting tumours—10% are bilateral, 10% extra-adrenal; 10% occur in children; 10% are malignant. May present with a huge array of symptoms and signs. ↑ BP may be sporadic or sustained. Suspect in patients who:

- Are young with ↑ BP
- Have very labile BP or sudden-onset hypertension
- Have ↑ BP and associated headaches, sweating, and/or palpitations
- Have other associated conditions (e.g. neurofibromatosis)

Check 24h urine catecholamine/metabolite levels (follow local laboratory protocol). If confirmed, or strong suspicion despite −ve test, refer for specialist opinion. Treatment is usually surgical if tumour is found.

Patient advice and support

Addison's Disease Self Help Group 🖱 www.addisons.org.uk
The Pituitary Foundation ☎ 0117 370 1320 🖱 www.pituitary.org.uk

Pituitary problems

Hypopituitarism ↓ production of all pituitary hormones (ACTH, growth hormone, FSH, LH, TSH, and prolactin). *Causes*:

- Iatrogenic—surgery/irradiation
- Infection—TB
- Tumour (may be non-secreting or secrete 1 pituitary hormone with ↓ secretion of the others)
- Sheehan's syndrome—pituitary necrosis following postpartum haemorrhage

Presentation

- Hypothyroidism
- Hypogonadism
- Anorexia
- Headache
- Depression
- Hair loss
- Hypotension
- Visual field defect

Management If suspected refer to neurology or endocrinology for further investigation and advice on treatment.

Pituitary tumours 10% intracranial tumours. Almost all are benign. Classified by histological type (chromophobic, acidophilic, or basophilic) or by the hormone secreted:

- No hormone (30%)
- Prolactin (35%)
- Growth hormone (20%)
- ACTH (7%)
- Prolactin and growth hormone (7%)
- LH, FSH, and TSH (1%)

Presentation Present with symptoms caused by:

- Local pressure—bilateral hemianopia, cranial nerve palsies, headache
- Hormone secretion, and/or
- Hypopituitarism (see earlier in this topic)

Management If suspected, refer for specialist management.

Pituitary apoplexy Rapid expansion of a pituitary tumour due to infarction or haemorrhage. Suspect if sudden onset of headache in a patient with a known pituitary tumour. Admit as a medical/neurosurgical emergency.

Craniopharyngioma Tumour originating from Rathke's pouch. 50% present as children with local pressure effects (see 'Pituitary tumours'). Refer as for pituitary tumours.

Hyperprolactinaemia The most common pituitary disorder resulting from pituitary adenoma (prolactinoma).

Presentation Tends to present earlier in ♀ than ♂. Symptoms are due to pressure effects or ↑ prolactin. Symptoms of ↑ prolactin:

- ♀: loss of libido, weight gain, apathy, vaginal dryness, menstrual disturbance, infertility, galactorrhoea
- ♂: impotence, ↓ facial hair

Investigation Check basal plasma prolactin (ask the laboratory for conditions under which they would like the sample taken).

Other causes of ↑ prolactin

- Pregnancy
- Breastfeeding
- Stress
- Sleep
- Hypothyroidism
- Drugs—phenothiazines, metoclopramide, domperidone, SSRIs, methyldopa, oestrogens
- Chronic kidney disease
- Sarcoidosis

Management Exclude other causes of ↑ prolactin. If no obvious other cause, suspect prolactin-secreting pituitary adenoma and refer for specialist opinion.

Acromegaly Rare condition due to a growth hormone secreting pituitary tumour. *Typical age at presentation*: 30–50y.

Presentation

- Local pressure symptoms
- Changes in appearance—coarse oily skin; change in facial appearance with coarsening of features; ↑ foot size; ↑ teeth spacing
- Other effects—deepening of voice, sweating, paraesthesiae, proximal muscle weakness, progressive heart failure, goitre
- Complications—DM, ↑ BP, cardiomyopathy, large bowel tumours

Investigation and management Check growth hormone levels. Refer to endocrinology.

Diabetes insipidus (DI) Caused by impaired water resorption by the kidney. 2 mechanisms:

- Cranial DI ↓ ADH secretion from the posterior pituitary. 50% idiopathic. *Other causes*: head injury, tumour, infection, sarcoidosis, vascular, inherited
- Nephrogenic DI Impaired response of the kidney to ADH. *Causes*: drugs (e.g. lithium), hypercalcaemia, pyelonephritis, hydronephrosis, pregnancy (rare)

Presentation Polydipsia, polyuria, dilute urine, dehydration.

Investigations U&E (Na^+ is often ↑), plasma and urine osmolality (plasma ↑, urine ↓—ratio >1). Specialist investigations (e.g. water deprivation test) confirm diagnosis.

Management Treat the cause.

- Cranial DI may be treated with intranasal desmopressin or surgery
- Nephrogenic DI may be treated with dietary restriction of protein and salt and/or bendroflumethiazide

Syndrome of inappropriate ADH (SIADH) Important cause of hyponatraemia. Diagnosis is made by finding a concentrated urine (sodium >20mmol/L) in the presence of hyponatraemia (<125mmol/L) or low plasma osmolality (<260mmol/kg), in the absence of hypovolaemia, oedema, or diuretics. Always requires specialist management. *Causes*:

- Malignancy e.g. small cell lung cancer; pancreas; lymphoma
- CNS disorders e.g. stroke; subdural haemorrhage; vasculitis (SLE)

Patient advice and support

The Pituitary Foundation ☎ 0117 370 1320 🖱 www.pituitary.org.uk

Chapter 12

Gastrointestinal medicine

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Assessment of abdominal pain

❗ Signs may be unclear in elderly patients, children, or those on steroids.

History Consider:

- Site of pain—Figure 12.1
- Onset: how long? How did it start? Change over time?
- Character of pain: colicky pain comes and goes in waves—results from GI obstruction, renal/biliary colic, gastroenteritis, or IBS
- Radiation
- Associated symptoms, e.g. nausea, vomiting, diarrhoea
- Timing/pattern, e.g. constant, colicky, relationship to food
- Exacerbating/relieving factors—including previous treatments tried
- Severity

Examination

- Temperature, pulse, BP, respiratory rate
- Anaemia or jaundice?
- Abdomen—site of pain (Figure 12.1); guarding/rebound tenderness?
- Rectal/vaginal examination as needed
- Consider urine dipstick/finger prick blood glucose testing as needed

Management Treat the cause (Table 12.1).

⚠ If acute or subacute onset severe pain, admit as a surgical emergency to hospital.

Table 12.1 Differential diagnosis of abdominal pain

Renal/urological
Renal colic
UTI
Pyelonephritis
Urinary retention/hydronephrosis
Torsion of the testis

Gynaecological
Ectopic pregnancy
Dysmenorrhoea
Endometriosis
Pelvic inflammatory disease
Ovarian torsion
Ovarian cyst—bleed/rupture
Gynaecological malignancy

Gastrointestinal
Surgical
Perforated bowel
Bowel obstruction
Intussusception
Strangulated hernia
Volvulus
Appendicitis
Meckel's diverticulum
Gallbladder disease
Pancreatitis
GI malignancy
Henoch Schönlein purpura
Medical
Gastritis
Peptic ulcer
Gastroenteritis
Crohn's/UC
IBS
Constipation
Diverticular disease
Liver disease

Other intra-abdominal
Sickle cell crisis
Ruptured spleen
Leaking/ruptured AAA
Mesenteric ischaemia
Mesenteric adenitis
Subphrenic abscess

Metabolic
DM—ketoacidosis
Porphyria
Addison's disease
Lead poisoning

Other extra-abdominal
Shingles/post-herpetic neuralgia
Spinal arthritis
Muscular pain
MI
CCF
Pneumonia

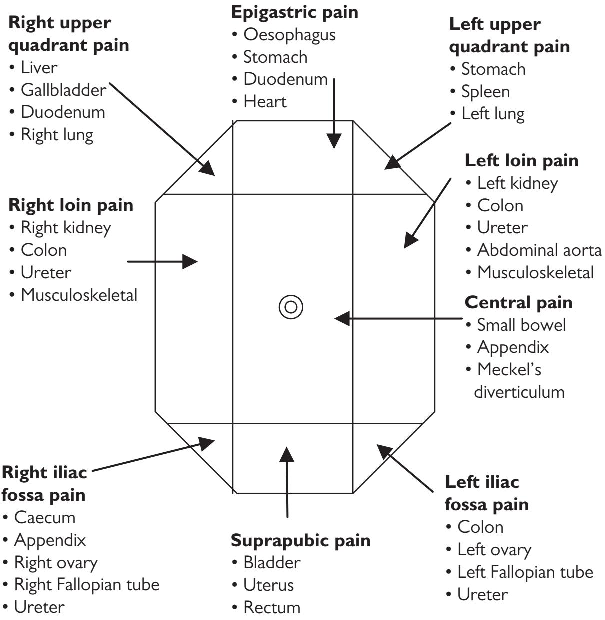
Right upper quadrant pain
• Liver
• Gallbladder
• Duodenum
• Right lung
Epigastric pain
• Oesophagus
• Stomach
• Duodenum
• Heart
Left upper quadrant pain
• Stomach
• Spleen
• Left lung
Left loin pain
• Left kidney
• Colon
• Ureter
• Abdominal aorta
• Musculoskeletal
Right loin pain
• Right kidney
• Colon
• Ureter
• Musculoskeletal
Central pain
• Small bowel
• Appendix
• Meckel's diverticulum
Right iliac fossa pain
• Caecum
• Appendix
• Right ovary
• Right Fallopian tube
• Ureter
Suprapubic pain
• Bladder
• Uterus
• Rectum
Left iliac fossa pain
• Colon
• Left ovary
• Left Fallopian tube
• Ureter

Figure 12.1 Location of pain and organs likely to be involved

Pelvic pain ➔ p. 690

Anal/perianal pain Treat the cause. *Consider:*

- Anal fissure
- Haemorrhoids/perianal haematoma (thrombosed pile)
- Perianal abscess
- Anal/perianal fistula
- Pilonidal sinus
- Skin infection (e.g. hidradenitis suppurativa)
- Functional pain (proctalgia fugax)
- Rectal/anal carcinoma

Tenesmus Sensation of incomplete rectal emptying following defecation—as if something has been left behind which cannot be passed. Common in irritable bowel syndrome. Can be also be caused by proctitis/inflammatory bowel disease and tumour.

Abdominal migraine or periodic syndrome Seen in children. Presents as stereotyped attacks in which nausea, vomiting, and headache accompany abdominal pain. Treat as for migraine. Some of these children develop classical migraine later.

Vomiting and diarrhoea

Most episodes of acute vomiting and diarrhoea are due to viral infection, short-lived (2–5d) and self-limiting.

Nausea Unpleasant symptom. The patient feels as if he/she might vomit. Most conditions which cause vomiting can also cause nausea.

Vomiting Common symptom. Causes—Table 12.2.

History

- Duration
- Ability to retain food and fluids/relationship to eating
- Nature of vomitus, e.g. presence of blood or 'coffee grounds'; bilious
- Sources of infection: food exposure (e.g. reheated rice, uncooked chicken); foreign travel; any contacts with similar symptoms?
- Other associated symptoms, e.g. fever, abdominal pain, diarrhoea
- Other illnesses, e.g. DM, Ménière's disease, migraine, cancer
- Medication, e.g. opioids, chemotherapy

Examination

- Assess hydration status—BP, pulse rate—dry mouth, ↓ skin turgor, sunken eyes, or sunken fontanelle (babies) are all late signs
- Abdomen—masses, distention, tenderness, bowel sounds
- For children—look for other sources of infection e.g. ENT, chest, UTI

Slimy stool Caused by overproduction of mucus in the large bowel. Almost always associated with colonic disease/irritable bowel syndrome. Investigate unless all other features are typical of IBS and age is <40y.

Diarrhoea Establish what the patient means by diarrhoea. Diarrhoea is the abnormal passage of loose or liquid stools. Causes—Table 12.2.

History

- Duration—termed 'chronic' if persists >4wk
- Nature of the diarrhoea—colour, consistency, blood/mucus
- Contact with anyone else with similar symptoms?
- Occupation and travel history
- Associated symptoms, e.g. fever, abdominal pain, vomiting, weight ↓
- Association with other factors (e.g. food intolerance, stress)
- Past medical history—surgery (especially ileal resection or cholecystectomy); pancreatic disease; systemic disease (e.g. DM, thyrotoxicosis)
- Family history—inflammatory bowel or coeliac disease; bowel cancer
- Alcohol consumption—high intake is associated with diarrhoea
- Medication e.g. antibiotics, regular medications (4% chronic diarrhoea)

Examination

- Assess hydration status—BP, pulse rate—dry mouth, ↓ skin turgor, sunken eyes, or sunken fontanelle (babies) are all late signs
- Abdomen—masses, distention, tenderness, bowel sounds, stool

Investigation Send a stool sample for M,C&S if any of the following:

- Fever
- Blood in stool
- Food worker
- Recent return from a tropical climate
- Immunocompromise
- Resident in an institution
- Persists >7d

Table 12.2 Causes of vomiting and diarrhoea

Vomiting	Diarrhoea
Physiological: e.g. posseting in babies *Travel/motion sickness* *GI infection*: e.g. viral gastroenteritis, food poisoning *Other infection* (particularly children): tonsillitis, otitis media *Other GI causes*: GI obstruction, pyloric stenosis, cow's milk protein allergy, 'acute abdomen' *CNS causes*: raised intracranial pressure, head injury, migraine, vertigo *Metabolic causes*: pregnancy, uraemia, ketoacidosis *Psychiatric causes*: anorexia, bulimia *Malignancy* *Drugs and toxins*: e.g. alcohol, opioids, cytotoxic agents	Acute diarrhoea • Dietary indiscretion • Infection, e.g. food poisoning, traveller's diarrhoea • Constipation with overflow • Pseudomembranous colitis—recent history of oral antibiotics • Onset of inflammatory bowel disease or • Other chronic diarrhoea Chronic diarrhoea Table 12.12, ➔ p. 379

Management of acute diarrhoea and/or vomiting

- Treat any identified cause
- Rehydration—encourage clear fluid intake (small amounts frequently) ± rehydration salts (use a commercial preparation e.g. Dioralyte®)
- Food—stick to a bland diet avoiding dairy products until symptoms have settled. Babies who are breastfed or have not been weaned should continue their normal milk
- If dehydrated and unable to replace fluids, e.g. diarrhoea with concomitant vomiting, or child/elderly person refusing to drink—admit

⚠ Never give children antidiarrhoeal agents.

❗ If no cause is found and diarrhoea lasts >4wk, or any atypical features consider referral for urgent investigation—➔ p. 378

Haematemesis ➔ p. 1061 **Gastroenteritis** ➔ p. 348

Faecal incontinence ➔ p. 380 **Factitious diarrhoea** ➔ p. 379

Chronic diarrhoea and malabsorption ➔ p. 378

Melaena or rectal bleeding ➔ p. 1061

- Some children may become cow's milk intolerant after a bout of gastroenteritis—➔ p. 867
- Think of haemolytic uraemic syndrome in any child with diarrhoea who passes blood in the stool

Further information

NICE (2009) Diarrhoea and vomiting in children under 5. 🖱 www.nice.org.uk/Guidance/CG84

Gastroenteritis and food poisoning

Ingestion of viruses, bacteria, or their toxins commonly causes diarrhoea and/or vomiting. ❗ Suspected food poisoning is a notifiable disease.

Prevention Handwashing after using the toilet; longer cooking and rewarming times; prompt consumption of food.

Presentation Common causes—Table 12.3.

- **History** Severity and duration of symptoms, food eaten and water drunk, time relationship between ingestion and symptoms, other affected contacts, recent foreign travel
- **Examination** Usually normal. Dehydration may prompt admission

Investigation and management See vomiting and diarrhoea—➔ p. 346. Advise fluid replacement. Only give antibiotics if recommended following stool culture (except *Giardia* diarrhoea—➔ p. 625).

Campylobacter Most common bacterial cause of infectious diarrhoea in the UK. 2 species (*C. jejuni* and *C. coli*) are responsible for most cases. Symptoms occur 2–5d after ingestion of infected food (usually milk or poultry). Malaise followed by abdominal pain and diarrhoea—often bloody. Rarely associated with arthritis. Usually clears spontaneously. If needed, treatment is with erythromycin or ciprofloxacin.

Salmonella Common cause of infectious diarrhoea. Usually ingested in infected meat, poultry, or eggs. *Symptoms:* vomiting, diarrhoea, abdominal pain, and fever—develop from 12h–2d after ingestion. Rarely associated with arthritis 2–3wk after acute infection. In <1% a carrier state develops. Only use antibiotics on microbiologist advice.

Escherichia coli Many different strains of *E.coli* cause diarrhoea via a variety of mechanisms. In most cases, treatment is supportive with fluid replacement. Rarely, for enterohaemorrhagic strains, antibiotics may be recommended, but use is controversial as antibiotic treatment has been linked with haemolytic uraemic syndrome.

Cryptosporidium Protozoan causing diarrhoeal disease. Infections are usually spread in water. Responsible for ~5% of all gastroenteritis in both industrialized and developing countries. Presents with profuse watery diarrhoea, abdominal cramp ± nausea, anorexia, fever, and malaise. *Treatment is* supportive. Usually symptoms last 1–2wk (rarely >1mo). Immunocompromised patients develop profuse intractable diarrhoea which is difficult to clear and may continue intermittently for life.

Norovirus ('winter vomiting virus') Most common cause of infectious gastroenteritis in the UK—particularly in communal settings, e.g. schools, hospitals. Illness is generally mild and lasts 2–3d. There are no long-term effects. Infections can occur at any age because immunity is not long-lasting. Scrupulous hygiene is needed to contain outbreaks.

Further information

Health Protection Infectious diseases: gastrointestinal infections. 🖱 www.gov.uk/topic/health-protection/infectious-diseases

Table 12.3 Common causes of gastroenteritis in the UK

Organism/source	Incubation	Symptoms					Food
		D	V	P	F	O	
B. cereus	1–5h	✓	✓				Rice
Campylobacter	48h–5d	✓		✓	✓	Blood in stool	Milk, poultry
C. botulinum	12–36h		✓			Paralysis	Canned food
C. perfringens	6–24h	✓		✓			Meat
E. coli	12–72h	✓		✓	✓	Blood in stool	Food, water
Salmonella spp.	12–48h	✓	✓	✓	✓		Meat, eggs, poultry
ShigellaND	48–72h	✓		✓	✓	Blood in stool	Any food
Staph. aureus	1–6h	✓	✓	✓		↓ BP	Meat
V. para-haemolyticus	12–24h	✓	✓	✓			Fish
Y. enterocolitica	24–36h	✓		✓	✓		Milk, water
Giardia lamblia	1–4wk	✓					Water
Entamoeba histolytica	1–4wk	✓		✓	✓	Blood in stool	Food, water
Cryptosporidium	4–12d	✓		✓	✓		Water
Listeria						Flu-like illness, pneumonia, miscarriage	Milk products, pâtés, raw vegetables
Norovirus	24–48h	✓	✓		✓	Malaise	Food, water
Rotavirus	1–7d	✓	✓		✓	Malaise	Food, water
Mushrooms	15min–24h	✓	✓	✓		Fits, coma, renal/liver failure	
Scombrotoxin	10–60min	✓				Flushes, erythema	Fish
Heavy metals, e.g. zinc	5min–2h		✓	✓			

D = diarrhoea; V = vomiting; P = abdominal pain; F = fever; O = other.

Rotavirus Most common cause of gastroenteritis in children. Most are immune by 5y. Presents with malaise, abdominal pain, diarrhoea, and vomiting. Common cause of hospital admission. Treatment is supportive. Babies in the UK are offered rotavirus vaccination within the childhood immunization programme (➔ p. 619).

❗ Some children may become cow's milk intolerant after a bout of gastroenteritis—➔ p. 867.

Constipation

3 million GP consultations/y in the UK result from constipation. Differentiate normal stools a few days apart (normal, needs no treatment) and infrequent hard stools (suggests constipation).

Definition 2 or more of the following for ≥3mo:

- Straining at defecation ≤25% of the time
- A sensation of incomplete evacuation ≥25% of the time
- ≤2 bowel movements/wk
- Lumpy and/or hard stools ≥25% of the time

❗ Most patients consulting in general practice do not meet these criteria.

Children with constipation ➔ p. 866

Young patients <40y with lone constipation ♀:♂ ≈9:1. Establish symptoms—constipation is usually longstanding in this group. Include drug and diet history. Ask about health beliefs—80% believe their bowels should open daily. Explore concerns about underlying disease. If long-standing ask why the patient is consulting now. Examine the abdomen. Investigate if symptoms/signs suggestive of organic disease (Table 12.4).

Management Treat organic causes. Otherwise:

- Give lifestyle advice—↑ fluid intake to ≤2L/d (8–10 cups); avoid alcohol; ↑ exercise if possible; add fibre to diet (↑ fruit/vegetables, eat wholegrain foods, add bran/oats); open bowel when needed
- If lifestyle advice fails and symptoms cause distress, start a bulk-forming laxative, e.g. ispaghula husk (avoid in opioid constipation). If this fails, try an osmotic laxative, e.g. a macrogol or MgOH 15mL bd
- If an osmotic laxative fails, try a short course of stimulant laxative, e.g. senna 1–2 tablets at 5 p.m. either alone or in combination with an osmotic laxative. Long-term use of some stimulant laxatives is reported to cause cathartic atonic colon. Although this is unlikely in young, fit patients only use short courses or use intermittently, e.g. twice weekly
- If still constipated, specialist referral is warranted

Table 12.4 Organic causes of constipation

Colonic disease	• Carcinoma • Diverticular disease • Crohn's disease	• Stricture • Intussusception • Volvulus
Anorectal disease	• Anterior mucosal prolapse • Distal proctitis	• Anal fissure • Perianal abscess
Pelvic disease	• Ovarian tumour • Uterine tumour	• Endometriosis
Endocrine/metabolic disorders	• Hypercalcaemia • Hypothyroidism	• DM with autonomic neuropathy
Drugs	• Opioids • Antacids containing calcium or aluminium • Antidepressants • Iron	• Antiparkinsonian drugs • Anticholinergics • Anticonvulsants • Antihistamines • Calcium antagonists
Other	• Pregnancy • Immobility	• Poor fluid intake

Irritable bowel syndrome (IBS) with constipation 20% develop symptoms of IBS in their lifetime (➔ p. 388). Constipation is the predominant symptom in 30% but other symptoms are usually present. Establish symptoms. Examine the abdomen. Investigation includes FBC, CRP, and TTG to exclude organic causes (➔ p. 388).

Management If <40y, examination and investigations are normal, and fulfils IBS criteria (➔ p. 388), manage as for young patients with lone constipation but avoid osmotic laxatives as they make bloating worse.

Constipation in the over 40s Any sustained change in bowel habit for >6wk should be taken seriously and investigated if appropriate. Establish symptoms and onset. Specifically ask about tenesmus, blood in stool, abdominal pain, and diarrhoea. Check current medication. Examine the abdomen for masses and hepatomegaly. Rectal examination is essential to exclude low rectal or anal carcinoma and detect faecal impaction.

Management

- Check FBC, ESR, renal function, LFTs, TFTs, and serum glucose
- Image the lower bowel by colonoscopy or CT colography if new symptoms that persist >6wk
- Treat any reversible, underlying organic cause—Table 12.4
- Give lifestyle advice (see management of constipation <40y, ➔ p. 350)
- Treat symptomatically if no cause is found/cause is untreatable
- Laxatives—consider a bulk-forming laxative (e.g. ispaghula) or osmotic (e.g. magnesium hydroxide, macrogol,) ± a stimulant laxative (e.g. senna). Titrate dose to response
- Long-term use of stimulant laxatives including co-danthrusate is acceptable in the very elderly. Otherwise use prn or intermittently
- If oral laxatives are ineffective, consider adding rectal measures. If soft stool, try bisacodyl suppositories (❗ must come into direct contact with rectum); if hard stools, try glycerol suppositories (act in 1–6h)
- If still not cleared/faecal impaction—refer to the district nurse for lubricant ± high phosphate (stimulant) enema (acts in ~2 min)
- Once constipation has been cleared, leave the patient with clear instructions about what to do if symptoms recur

⚠ **High-risk patients** e.g. patients on opioids; those who are immobile or have medical conditions which predispose them to constipation. Pre-empt constipation by putting high-risk patients on regular aperients.

Occult presentations of constipation are common in the elderly and include:

- Confusion
- Urinary retention
- Abdominal pain
- Overflow diarrhoea
- Loss of appetite and nausea

Further information

NICE CKS (2017) Constipation 🔗 https://cks.nice.org.uk/constipation

Other abdominal symptoms and signs

Dyspepsia ➔ p. 354

Abdominal distention Consider abdominal/pelvic masses and:

- Fluid—ascites or full bladder
- Fat
- Faeces
- Flatus—intestinal obstruction; air swallowing
- Fetus
- Food, e.g. malabsorption

Abdominal masses Distinguished from pelvic masses by the ability to get beneath them. *Causes:* malignancy—any intra-abdominal organ or kidney; stool; abdominal aortic aneurysm; hepato- and/or splenomegaly; appendix mass/abscess; Crohn's mass; lymph nodes or TB mass. ❗ A hernia may present as a mass in abdominal wall/groin lump—➔ p. 364.

Pelvic masses *Causes*: fetus; full bladder; fibroids; gynaecological malignancy; bladder cancer.

Splenomegaly *Causes*:

- Haematological Lymphoma, leukaemia, myeloproliferative disorders, sickle cell disease (children usually), thalassaemia
- Inflammatory RA or Sjögren's syndrome, sarcoid, amyloid
- Infection Glandular fever, malaria, SBE, TB, leishmaniasis

Hepatomegaly *Causes*:

- Apparent Riedel's lobe, low-lying diaphragm
- Tumours Secondary (most common), primary
- Venous congestion Heart failure, hepatic vein occlusion
- Haematological Leukaemia, lymphoma, myeloproliferative disorders, sickle cell disease
- Biliary obstruction Particularly extrahepatic
- Inflammation Hepatitis, abscess, schistosomiasis
- Metabolic Fatty liver, amyloid, glycogen storage disease
- Cysts Polycystic liver, hydatid

Ascites Free fluid in the peritoneal cavity. *Signs*: abdominal distention, shifting dullness to percussion, fluid thrill. *Causes*: malignancy—any intra-abdominal organ, ovary, or kidney; hypoproteinaemia, e.g. nephrotic syndrome; right heart failure; portal hypertension.

Fistula Abnormal communication between 1 organ and another—usually due to cancer, or complication of surgery. Presentation—Table 12.5. Refer urgently if suspected.

Table 12.5 Presentation of fistula

Connection	Presentation
Bowel → skin	Faecal discharge through surgical wound
Bladder/ureters → skin	Clear, watery discharge which smells of urine
Bowel → vagina	Feculent material in vagina
Bladder → vagina	Leakage of urine per vaginum
Bowel → bladder	Air or feculent material in urine; recurrent UTI

⚠ Referral for suspected GI cancer[N]

Urgent referral (to be seen in <2wk)

To a team specializing in upper GI cancer if:

- Upper abdominal mass consistent with stomach/pancreatic cancer
- Aged ≥40y + jaundice

To a team specializing in colorectal cancer if:

- Any age + anal ulceration
- Any age + anal, rectal, or abdominal mass
- Any age + rectal bleeding + unexplained abdominal pain, change in bowel habit, weight loss, or iron deficiency anaemia
- Aged ≥40y + unexplained weight loss + abdominal pain, or
- Aged ≥50y + unexplained rectal bleeding, or
- Aged ≥60y + iron-deficiency anaemia or persistent change in bowel habit or faecal occult blood +ve

For upper GI endoscopy if:

- Dysphagia
- Aged ≥55y + weight loss + upper abdominal pain, reflux, and/or dyspepsia

For direct access CT (or USS if CT not available) if: ≥60y + weight loss AND ≥1 of:

- Diarrhoea
- Back pain
- Abdominal pain
- Nausea/vomiting
- Constipation
- New-onset diabetes

For direct access USS if: upper abdominal mass consistent with enlarged liver or gallbladder.

Non-urgent referral

For upper GI endoscopy if:

- Haematemesis, or
- Aged ≥55y with:
  - Treatment-resistant dyspepsia, or
  - Upper abdominal pain and ↓ Hb, or
  - ↑ platelet count + nausea/vomiting, weight ↓, reflux, dyspepsia, and/or upper abdominal pain, or
  - Nausea/vomiting + weight loss, reflux, dyspepsia, and/or upper abdominal pain

❗ *H. pylori* status should not affect the decision to refer for suspected cancer. Consider checking a FBC to exclude iron deficiency anaemia in all patients presenting with new-onset dyspepsia.

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Dyspepsia and *H. pylori*

In any year, up to 40% of the adult population suffer from dyspepsia—1:10 seek their GP's advice; ~10% of these are referred for endoscopy.

Causes

- Gastro-oesophageal reflux disease (GORD)—15–25% ➔ p. 358
- Peptic ulcer (PU)—15–25% ➔ p. 360
- Stomach cancer—2% ➔ p. 362
- 60% are classified as *non-ulcer dyspepsia* (NUD, 'functional' dyspepsia)—manage as for uninvestigated dyspepsia (Figure 12.2)
- *Rarer causes*—oesophagitis from swallowed corrosives, oesophageal infection (especially in the immunocompromised)

Differential diagnosis Cardiac pain (difficult to distinguish), gallstone pain, pancreatitis, bile reflux.

Presentation Common symptoms include retrosternal or epigastric pain, fullness, bloating, wind, heartburn, nausea, and vomiting. Examination is usually normal though there may be epigastric tenderness. Check for clinical anaemia, epigastric mass/hepatomegaly, and LNs in the neck.

Management Figure 12.2

Lifestyle advice Give advice on healthy eating, weight ↓, and smoking cessation. Avoid precipitating factors, e.g. alcohol, coffee, chocolate, fatty foods. Raising the head of the bed and having a main meal well before going to bed may help. Promote continued use of antacids/alginates.

Helicobacter pylori Infection is associated with:

- GI disease—peptic ulcer disease; gastric cancer; non-ulcer dyspepsia; oesophagitis
- Non-GI disease—ranging from cardiovascular disease and haematological malignancy to cot death

Testing for H. pylori^{N} 'Test and treat' all patients with dyspepsia who do not meet referral criteria (Figure 12.2). Choice of test is limited by availability, ease of access, and cost. Community options are: serology, urea breath test, and faecal antigen test. A 2wk washout period following proton pump inhibitor (PPI) use is necessary before testing for *H. pylori* with a breath test or a stool antigen test.

Eradication^{N} Clears 80–85% *H. pylori* infections. First-line options:

- PAC$_{500}$ regimen Full-dose PPI (e.g. omeprazole 20mg bd) + amoxicillin 1g bd + clarithromycin 500mg bd for 1wk *or*
- PMC$_{250}$ regimen Full-dose PPI (e.g. omeprazole 20mg bd) + metronidazole 400mg bd + clarithromycin 250mg bd for 1wk

For more first-line options and second-line options see NICE guidelines.

❗ Do not retest even if dyspepsia remains unless there is a strong clinical need. Retest if needed using a urea breath test.

Further information

NICE (2014) Gastro-oesophageal reflux disease and dyspepsia in adults: investigation and management. 🖱 www.nice.org.uk/Guidance/CG184

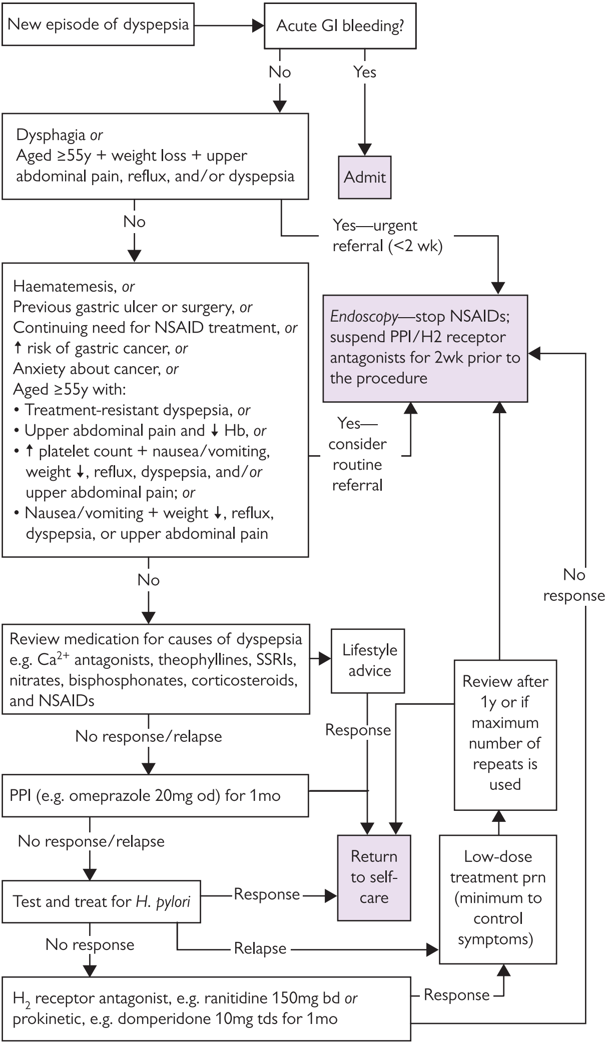
New episode of dyspepsia
Acute GI bleeding?
No
Yes
Admit
Dysphagia *or*
Aged ≥55y + weight loss + upper abdominal pain, reflux, and/or dyspepsia
No
Yes—urgent referral (<2 wk)
Haematemesis, *or*
Previous gastric ulcer or surgery, *or*
Continuing need for NSAID treatment, *or*
↑ risk of gastric cancer, *or*
Anxiety about cancer, *or*
Aged ≥55y with:
• Treatment-resistant dyspepsia, *or*
• Upper abdominal pain and ↓ Hb, *or*
• ↑ platelet count + nausea/vomiting, weight ↓, reflux, dyspepsia, and/*or* upper abdominal pain; *or*
• Nausea/vomiting + weight ↓, reflux, dyspepsia, or upper abdominal pain
Endoscopy—stop NSAIDs; suspend PPI/H2 receptor antagonists for 2wk prior to the procedure
Yes—consider routine referral
No
No response
Review medication for causes of dyspepsia e.g. Ca^{2+} antagonists, theophyllines, SSRIs, nitrates, bisphosphonates, corticosteroids, and NSAIDs
Lifestyle advice
Response
Review after 1y or if maximum number of repeats is used
No response/relapse
PPI (e.g. omeprazole 20mg od) for 1mo
No response/relapse
Return to self-care
Low-dose treatment prn (minimum to control symptoms)
Test and treat for *H. pylori*
Response
No response
Relapse
H_2 receptor antagonist, e.g. ranitidine 150mg bd *or* prokinetic, e.g. domperidone 10mg tds for 1mo
Response

Figure 12.2 Algorithm for management of uninvestigated dyspepsia in general practice

Oesophageal conditions

Oesophagitis Common condition. Reflux of acid from the stomach to the oesophagus causes mucosal damage resulting in inflammation and ulceration. *Other causes*: drugs (e.g. NSAIDs); infection (e.g. CMV, HSV, candida—especially in the immunocompromised); ingestion of caustic substances.

Management Treat reflux-induced oesophagitis as for GORD—➔ p. 358. Otherwise treat the cause.

Barrett's oesophagus ➔ p. 359.

Chronic benign stricture Recurrent oesophagitis (e.g. 2° to GORD, NSAIDs, K^+ preparations) scars the oesophagus resulting in stricture formation. Most common in elderly ♀.

Presentation Long history of reflux with more recent dysphagia. If obstruction is severe undigested food may be regurgitated immediately after swallowing. May be associated with night-time coughing paroxysms due to aspiration of gastric contents into the chest. Examination is usually normal.

Management Refer for urgent endoscopy to confirm diagnosis and exclude carcinoma. Treatment is by endoscopic dilatation of the stricture.

Carcinoma of the oesophagus ➔ p. 362

Presbyoesophagus Common among the elderly. Intermittent sensation that food is getting stuck—usually at the back of the throat. Examination is normal as is endoscopy. Barium swallow or oesophageal motility studies may reveal oesophageal spasm. Reassure.

Globus pharyngis (or hystericus) Sensation of a lump in the throat without difficulty swallowing is common. It may indicate anxiety. Reassure if no organic signs and treat any dyspepsia. If not responding refer to ENT for exclusion of an organic cause.

Eosinophilic oesophagitis Chronic, allergic oesophagitis. ♂ > ♀. More common if history of atopy. FH in 7%. Most present aged <40y with oesophageal dysfunction (dysphagia, food impaction, vomiting, regurgitation, heartburn, abdominal pain). In children may present with failure to thrive or feeding intolerance. If suspected, refer for endoscopy. Oesophageal biopsy shows characteristic eosinophilic infiltration. Treatment is with a PPI for 8wk to exclude GORD; if endoscopic changes persist, treatment is with an exclusion or elemental diet and/or topical glucocorticoids. Oesophageal dilatation may be needed in severe cases.

Oesophageal achalasia Failure of relaxation of the circular muscles at the distal oesophagus. *Peak incidence*: 30–40y; ♀ slightly > ♂.

Presentation Gradual onset of dysphagia over years accompanied by regurgitation of stagnant food and foul belching. Night-time coughing fits are due to aspiration which can result in recurrent chest infections. Examination is usually normal although there may be signs of aspiration pneumonia.

Management CXR to exclude aspiration pneumonia; endoscopy confirms diagnosis. Refer for surgery.

Plummer–Vinson syndrome Iron deficiency anaemia + dysphagia due to a post-cricoid web in the oesophagus. ♀ > ♂. *Peak incidence*: 40–50y. Presents with high dysphagia with food sticking in the back of the throat ± retching/choking sensation. This is a premalignant condition so refer for biopsy and dilatation of pharyngeal web; replace iron.

Pharyngeal pouch Pulsion diverticulum of the pharyngeal mucosa through Killian's dehiscence (area of weakness between the 2 parts of the inferior pharyngeal constrictor). ♂ > ♀; ↑ with age. Usually develops posteriorly then protrudes to 1 side—L > R. As the pouch gets larger, the oesophagus is displaced laterally.

Presentation Dysphagia—the first mouthful is swallowed easily then fills the pouch which makes further swallowing difficult. Accompanied by regurgitation of food from the pouch ± symptoms of aspiration (night-time coughing, recurrent chest infection). A swelling is palpable in the neck in ⅔ of cases.

Management Refer for further investigation. Diagnosis is confirmed with endoscopy/barium swallow. Treatment is surgical.

Oesophageal varices Result from portal hypertension (➔ p. 397) and can bleed massively—admit as a 'blue light' emergency if bleeding.

Impacted oesophageal foreign body Usually the patient notices something has stuck resulting in pain, difficulty swallowing ± retching. If suspected refer immediately to A&E for further investigation ± removal of the foreign body.

Oesophageal perforation Rare—usually a complication of endoscopy. Less commonly due to violent vomiting. The patient becomes very distressed with pain relating to the site of perforation which is worse on swallowing. Examination reveals tachycardia, shock ± pyrexia ± breathlessness ± surgical emphysema in neck. Admit as a surgical emergency.

Oesophageal atresia and/or tracheo-oesophageal fistula

1:2500 live births. 5% have oesophageal atresia alone; 5% tracheo-oesophageal fistula (TOF) alone; the remainder have both. Risk factors for sudden infant death syndrome.

Presentation

- Antenatal: at routine USS or following investigation of polyhydramnios
- Postnatal: cough or breathing difficulties in a newborn infant, choking on the first feed, inability to swallow saliva → bubbling of fluid from the mouth developing soon after birth
- Later in childhood: 'H-type' fistulas where there is no atresia but just a fistula may present late with recurrent chest infections

Management Diagnosis is confirmed with X-ray. Treatment is surgical. Postoperatively children may have a barking cough ('*TOF cough*') and/or dysphagia—both settle before 2y.

Gastro-oesophageal reflux and gastritis

Gastro-oesophageal reflux disease (GORD) Caused by retrograde flow of gastric contents through an incompetent gastro-oesophageal junction. It affects ~5% of the adult population.

Risk factors

- Smoking
- Alcohol
- Coffee
- Fatty food
- Big meals
- Obesity
- Hiatus hernia
- Tight clothes
- Pregnancy
- Systemic sclerosis
- Drugs (NSAIDs, TCAs, SSRIs, iron supplements, anticholinergics, nitrates, alendronic acid)
- Surgery for achalasia

Conditions caused by GORD

- Oesophagitis (defined by mucosal breaks) ± oesophageal ulcer
- Benign oesophageal stricture—➔ p. 356
- Intestinal metaplasia: Barrett's oesophagus
- Oesophageal haemorrhage
- Anaemia

Presentation

- Heartburn: most common symptom. Burning retrosternal or epigastric pain which worsens on bending, stooping or lying, and with hot drinks. Relieved by antacids
- Other symptoms:
  - Waterbrash—mouth fills with saliva
  - Reflux of acid into the mouth—especially on lying flat
  - Nausea and vomiting
  - Nocturnal cough/wheeze due to aspiration of refluxed stomach contents
- Examination: usually normal. Check for clinical anaemia, epigastric mass/hepatomegaly and LNs in the neck

Investigation Endoscopy if indicated—see Figure 12.2, ➔ p. 355

❗ Symptoms are poorly correlated with endoscopic findings. Reflux may remain silent in patients with Barrett's oesophagus but heartburn can severely affect quality of life of patients with −ve endoscopy results.

Initial management^{N}

- In all cases give lifestyle advice (➔ p. 354)
- If diagnosis is clinical (i.e. patient presents with 'reflux-like' symptoms), treat as for uninvestigated dyspepsia (Figure 12.2, ➔ p. 355)
- If reflux confirmed on endoscopy, offer treatment with a PPI (e.g. omeprazole 20mg od) for 1–2mo
- If oesophagitis at endoscopy and still symptomatic on standard dose PPI, double the PPI dose (e.g. omeprazole 20mg bd) for a further 1mo
- If inadequate response to PPI, try an H_2 receptor antagonist (e.g. ranitidine 150mg bd) and/or add a prokinetic (e.g. metoclopramide 10mg tds) for 1mo

Long-term management of endoscopic/barium-confirmed GORD^{N}

- Patients who have had dilatation of an oesophageal stricture should remain on long-term full-dose PPI therapy

- For all other patients, if symptoms recur following initial treatment, offer a PPI at the lowest dose possible to control symptoms, with a limited number of repeat prescriptions. Discuss using the treatment on an as-required basis to manage symptoms
- Refer for consideration of surgery if quality of life remains significantly impaired despite optimal treatment or if patient does not wish to continue PPI/H_2 receptor antagonist therapy long term. Surgery of any type is >90% successful although results may deteriorate with time

Hiatus hernia Common (30% of over 50s); 50% have GORD. Obesity is a risk factor. The proximal stomach herniates through the diaphragmatic hiatus into the thorax.

- 80% have a 'sliding' hiatus hernia where the gastro-oesophageal junction slides into the chest
- 20% have a 'rolling' hernia where a bulge of stomach herniates into the chest alongside the oesophagus. The gastro-oesophageal junction remains in the abdomen

Management Treat as for GORD.

Barrett's oesophagus Usually found incidentally at endoscopy for symptoms of GORD and caused by chronic GORD. The squamous mucosa of the oesophagus undergoes metaplastic change and the squamocolumnar junction appears to migrate away from the stomach. The length affected varies. Associated with ↑ risk of adenocarcinoma of the oesophagus. Consider referral for endoscopic surveillance[N] if:

- Dysplasia
- Other risk factors—♂, older ager, ↑ length of Barrett's segment,

Treatment is with long-term PPIs (e.g. omeprazole 20–40mg od) ± laser therapy ± resection.

Acute gastritis Mucosal inflammation of the stomach with no ulcer.

- Type A—affects the entire stomach; associated with pernicious anaemia; premalignant
- Type B—affects antrum ± duodenum; associated with *H. pylori*
- Type C—due to irritants e.g. NSAIDs, alcohol, bile reflux

Presentation and investigation Dyspepsia—see ➔ p. 354

Management

- Treat the cause if possible (e.g. *H. pylori* eradication; ↓ alcohol)
- Acid suppression—H_2 receptor antagonist (e.g. ranitidine, nizatidine) or PPI for 4–8wk
- Re-endoscope to confirm healing

Complications Haemorrhage, gastric atrophy ± gastric cancer (type A only).

Further information

NICE (2014) Gastro-oesophageal reflux disease and dyspepsia in adults: investigation and management. 🖱 www.nice.org.uk/Guidance/CG184

Peptic ulceration

Peptic ulceration (PU) is a term which includes both gastric and duodenal ulceration. Most patients present with dyspepsia (➔ p. 354). Specific features of gastric and duodenal ulcers are listed in Table 12.6.

Management

For patients not taking NSAIDs

- Eradicate *H. pylori* if present—➔ p. 354—speeds ulcer healing and ↓ relapse; confirm eradication with a urea breath test (duodenal ulcer) or repeat endoscopy (gastric ulcer), and retreat if still present
- If *H. pylori* negative Treat with full-dose PPI (e.g. omeprazole 20mg od) for 1–2mo. If gastric ulcer, re-endoscope to check ulcer is healed

For patients taking NSAIDs

- Stop NSAIDs where possible. If not possible, consider changing to a safer alternative (e.g. paracetamol, ↓ dose of NSAID, COX2 selective NSAID) and adding gastric protection with a PPI or misoprostol
- Offer full-dose PPI or H_2 receptor antagonist (H_2RA) therapy for 2mo and, if *H. pylori* is present, subsequently offer eradication therapy
- Check eradication with repeat endoscopy (gastric ulcer) or urea breath test (duodenal ulcer)

For all patients

- Lifestyle measures Avoid foods (or alcohol) which exacerbate symptoms; eat little and often; avoid eating <3h before bed. Stop smoking
- If symptoms recur following initial treatment Offer a PPI at lowest dose to control symptoms, with a limited number of repeat prescriptions. Discuss using the treatment on a prn basis
- Offer H_2RA therapy If there is an inadequate response to a PPI
- In patients with unhealed ulcer or continuing symptoms despite adequate treatment Exclude non-adherence, malignancy, failure to detect *H. pylori*, inadvertent NSAID use, other ulcer-inducing medication, and rare causes, e.g. Zollinger–Ellison syndrome, Crohn's disease
- Once symptoms are controlled Review at least annually to discuss symptom control, lifestyle advice, and medication
- Refer If gastric ulcer fails to heal or if symptoms do not respond to medical treatment. Possible surgical procedures include: gastrectomy, vagotomy and drainage procedure; highly selective vagotomy

Zollinger–Ellison syndrome Association of peptic ulcer with a gastrin-secreting pancreatic (rarely duodenal) adenoma—50–60% are malignant, 10% are multiple, and 30% are associated with multiple endocrine neoplasia (MEN I). *Incidence*: 0.1% of patients with duodenal ulcer disease. Suspect in those with multiple peptic ulcers resistant to drugs, particularly if associated with diarrhoea ± steatorrhoea or a family history of peptic ulcers (or islet cell, pituitary, or parathyroid adenomas). Refer for further investigation. Treatment is with PPIs (e.g. omeprazole 10–60mg bd) ± surgery.

Table 12.6 Features of gastric and duodenal ulcers

	Gastric ulcer (GU)	Duodenal ulcer (DU)
Population	Typically affects middle aged/ elderly ♂	Typically affects young–middle-aged ♂ although can affect any adult. ♂ > ♀
Risk factors	*H. pylori* (70–90%) NSAID use (↑ risk ×3–4) Delayed gastric emptying Reflux from the duodenum (↑ by smoking)	*H. pylori* (>90%) NSAID use Gastric hyperacidity Rapid gastric emptying Smoking Stress (💣)
Presentation	May be asymptomatic Epigastric pain worsened by food and helped by antacids or lying flat ± weight loss With complications (see below)	May be asymptomatic or spontaneously relapse and remit Epigastric pain typically relieved by food and worse at night ± weight ↑ ± waterbrash (saliva fills the mouth) With complications (see below)
Examination	In uncomplicated gastric ulceration, examination is usually normal though there may be epigastric/left upper quadrant tenderness	In uncomplicated duodenal ulceration, examination is usually normal though there may be epigastric tenderness
Investigation	As for dyspepsia (➔ p. 355)	
Complications	*Bleeding:* acute GI bleeding—➔ p. 1061; iron deficiency anaemia—➔ p. 638 *Perforated peptic ulcer:* DU > GU; GUs may perforate posteriorly into the lesser sac; DUs usually perforate anteriorly into the peritoneal cavity. There may not be a past history of indigestion. Presents with sudden-onset severe epigastric pain which rapidly becomes generalized. When a GU perforates into the lesser sac symptoms may remain localized or be confined to the right side of the abdomen. Examination: generalized peritonism with 'board-like rigidity'. Management: acute surgical admission *Pyloric stenosis in adults:* duodenal stenosis 2° to scarring from a chronic DU. Characterized by copious vomiting of food 1–2 days old. There may not be a past history of indigestion. Examination: if prolonged vomiting may be evidence of dehydration ± weight ↓. Succussion splash may be audible. Management: surgical referral for confirmation of diagnosis and surgical relief	

Further information

NICE (2014) Gastro-oesophageal reflux disease and dyspepsia in adults: investigation and management. 🖱 www.nice.org.uk/Guidance/CG184

Gastro-oesophageal malignancy

Carcinoma of the oesophagus Common cancer accounting for 7500 deaths/y in the UK. Most common in patients >60y. Overall ♂:♀ ≈5:1. Usually presents late when prognosis is poor. 2 types:

- Squamous cell carcinoma (50%)—predominant form in upper ⅔ of the oesophagus
- Adenocarcinoma (50%)—predominant in lower ⅓ of the oesophagus. Incidence is ↑. ♂:♀ ≈5:1

Common risk factors

Squamous cell carcinoma:

- Smoking*
- Alcohol
- Low fruit/vegetable intake

Adenocarcinoma:

- Smoking*
- Obesity
- Low fruit/vegetable intake
- GORD—particularly Barrett's oesophagus (risk ↑ >30×—the longer the affected segment, the higher the risk)

* Risk ↓ to that of a non-smoker 10y after giving up.

Other risk factors

- Previous mediastinal radiotherapy (↑ ×2 for patients treated for breast cancer; ↑ × 20 for patients treated for Hodgkin's lymphoma)
- Plummer–Vinson (or Patterson–Kelly) syndrome—oesophageal web and iron deficiency anaemia
- Tylosis—rare, inherited disorder with hyperkeratosis of the palms—40% develop oesophageal cancer

Presentation Short history of rapidly progressive dysphagia affecting solids initially then solids and liquids ± weight loss ± regurgitation of food and fluids (may be bloodstained). Retrosternal pain is a late feature. Other symptoms include hoarseness and/or cough (due to aspiration or fistula formation). Examination may be normal. Look for evidence of recent weight loss, hepatomegaly, and cervical lymphadenopathy.

Management Refer for urgent endoscopy if suspected. Rapid-access dysphagia clinics are run in many areas. Specialist management involves resection (treatment of choice but only 1:3 patients are suitable), chemotherapy, radiotherapy, and/or palliation with a stenting tube. Tubes commonly become blocked. Good palliative care is essential—refer early (➲ p. 1011). Overall 8% 5y survival.

Stomach cancer Stomach cancer causes ~5000 deaths/y in the UK; 95% are adenocarcinomas. Disease affecting older people with 92% diagnosed >55y; ♂ > ♀ (5:3). Incidence has more than halved over the past 30y in the UK probably due to improved diet.

Other risk factors Include:

- Geography—common in Japan
- Blood group A
- *H. pylori* infection (not clear if eradication ↓ risk)
- Atrophic gastritis
- Pernicious anaemia
- Smoking
- Adenomatous polyps
- Social class
- Previous partial gastrectomy

Presentation Often non-specific. Presents with dyspepsia, weight ↓, anorexia or early satiety, vomiting, dysphagia, anaemia, and/or GI bleeding. Suspect in any patient >55y with recent-onset dyspepsia (within 1y) and/or other risk factors. Examination is usually normal until incurable. Look for epigastric mass, hepatomegaly, jaundice, ascites, enlarged supraclavicular LN (Virchow's node), acanthosis nigricans.

Management If suspected refer for urgent endoscopy. In early stages total/partial gastrectomy may be curative. Most present at later stage. Overall 5y survival is 15%.

Post-gastrectomy syndromes

Abdominal fullness A feeling of early satiety ± weight loss. Advise to take small, frequent meals.

Bilious vomiting Affects ~10% patients post-gastrectomy. Intermittent sudden attacks of bilious vomiting 15–30min after eating ± epigastric cramping pain relieved by vomiting. Usually settles spontaneously. Metoclopramide may be helpful in the interim. If symptoms are severe or fail to settle, request surgical review. Surgical bile diversion or stomach reconstruction may alleviate symptoms.

Dumping Abdominal distension, colic, and vasomotor disturbance (e.g. sweating, fainting) after meals. Affects 1–2% of gastrectomy patients (more common early after surgery—most settle within 6mo). 2 types:

- Early dumping Due to rapid gastric emptying. Starts immediately after a meal. *Consists of*: sweating, flushing, tachycardia, palpitations, epigastric fullness, and nausea. Occasionally there may be vomiting, diarrhoea, ± colicky abdominal pain. *Advise*: small, dry meals with restricted carbohydrate. Take drinks between meals. If severe, re-refer
- Late dumping Due to rapid gastric emptying → hyperglycaemia. The resultant hyperinsulinaemia causes a rebound hypoglycaemia. Starts 1–2h after meals. Consists of: faintness, sweating, tremor, and nausea. Advise patients to ↓ the sugar content of meals, rest for 1h after each meal, and take glucose if symptoms occur. If severe, re-refer

Diarrhoea post-gastrectomy 50% of patients who have had a truncal vagotomy or gastrectomy suffer some frequency of defecation; 5% require treatment. The diarrhoea is typically episodic and unpredictable. The exact mechanism is not clear. Treatment is with codeine phosphate or loperamide prn. Antibiotic treatment is occasionally successful—seek expert advice. Surgical measures are rarely necessary.

Anaemia Gastrectomy can result in both vitamin B_{12} deficiency and iron deficiency anaemia. Prophylactic vitamin B_{12} injections may be advised by the operating surgeon. Many advise iron supplements for life. An annual FBC to monitor for anaemia is advisable. Treat with iron/vitamin B_{12} supplements.

Stomach cancer Risk of stomach cancer is ↑ after partial gastrectomy (2× after 20y and 7× after 45y).

Advice and support for patients

Cancer Research UK ☎ 0808 800 4040 🖱 www.cancerresearchuk.org/about-cancer/stomach-cancer

Hernias

⚠ Irreducible hernia

- Most types of hernia may become irreducible
- It may be the first presentation of a hernia or a complication of a longstanding hernia
- If obstructed (incarcerated) or strangulated (blood supply to bowel contained within the hernia sac is compromised), the hernia is tender and there are symptoms/signs of small bowel obstruction

⚠ If you are unable to reduce a hernia, admit for surgical assessment.

Inguinal hernia Protuberance of peritoneal contents through the abdominal wall where it is weakened by the presence of the inguinal canal. Common condition (♂ > ♀) which can occur at any age.

Presentation Lump in the groin ± discomfort on straining/standing for any length of time. There may be a distinct precipitating event (e.g. heavy lifting). *Risk factors*: chronic cough (e.g. COPD), constipation, urinary obstruction, heavy lifting, ascites, previous abdominal surgery. *2 types*:

- Indirect (80%) Follow the course of the spermatic cord or round ligament down the inguinal canal through the internal inguinal ring (located at the mid-point of the inguinal ligament, 1.5cm above the femoral pulse) and sometimes out through the external inguinal ring into the scrotum/vulva
- Direct (20%) Pass through a defect in the abdominal wall into the inguinal canal. Rare in children and more common in the elderly

Differential diagnosis of groin lumps Table 12.7

Examination Examine the patient standing up. Look for a bulge in the groin above the line of the inguinal ligament. Unless incarcerated the lump should have a cough impulse. Check that you are able to reduce the hernia—sometimes it is easier if the patient lies down. Ask the patient to reduce the hernia if you cannot.

Management Small hernias often require no treatment. For larger hernias and smaller hernias that are symptomatic, consider referral for surgical repair. Various methods are used—all have a high level of success (<2% recurrence). Trusses can be useful for symptomatic hernias in elderly patients, those unfit for surgery, or while awaiting surgery (prescribe on FP10).

Inguinal hernias in children ➔ p. 868

Femoral hernia Less common than inguinal hernia. ♀ > ♂. The patient is usually elderly, although can occur at any age. Peritoneal contents protrude down the femoral canal. Risk of strangulation is high. Presents as a painful lump in the groin and/or small bowel obstruction.

Examination Rounded swelling medially in the groin and lateral to the pubic tubercle; if reducible a soft palpable lump remains after reduction.

Management Always refer for urgent surgical repair. Admit as surgical emergency if obstructed or irreducible.

Table 12.7 Differential diagnosis of groin lumps

Position relative to the skin	Groin lump	Position relative to the inguinal ligament	
		Above	Below
In the skin	Lipoma, fibroma, haemangioma, and other skin lumps	✓	✓
Deep to the skin	Femoral or inguinal lymph nodes	✓	✓
	Saphena varix of the femoral vein	✗	✓
	Femoral artery aneurysm	✗	✓
	Femoral hernia	✗	✓
	Inguinal hernia	✓	✗

Incisional hernia Breakdown of the muscle closure in an abdominal wound sometime after surgery. There may be a history of wound sepsis, haematoma, or breakdown. Presents with a bulge at the site of the operation scar ± discomfort.

Examination The hernia is usually visible when the patient stands—it can be made more obvious by asking the patient to cough or straight leg raise while lying flat. The margins of the muscular defect are palpable under the skin. Note whether fully reducible or not.

Management Often reassurance suffices. If obstructed/strangulated or causing discomfort, then refer for surgical assessment.

Umbilical hernia Most common in infants (➔ p. 868). In adults paraumbilical hernias, presenting as a bulge adjacent to the umbilicus, may occur due to weakness in the linea alba. ♀ > ♂. Refer adults for surgical assessment—usually repaired as risk of strangulation is high. Admit as a surgical emergency if obstructed/irreducible.

Epigastric hernia Midline hernia through a defect in the linea alba above the umbilicus. Never contains bowel. Usually symptomless though occasionally causes epigastric pain ± vomiting. *Examination*: epigastric mass with cough impulse. Refer for surgical repair.

Spigelian hernia A hernial sac protrudes lateral to the rectus sheath midway between umbilicus and pubic bone. Presents with discomfort ± vomiting. Refer for surgical repair.

Obturator hernia Hernia protrudes out from the pelvis through the obturator canal. Usually presents with strangulation ± pain referred to the knee. Admit for surgery.

Richter hernia A knuckle of the side wall of the gut gets caught in a hernia sac and becomes strangulated but the bowel is not obstructed. Presents with abdominal pain which rapidly becomes worse ± shock. Admit as for acute abdomen; diagnosis is usually made at surgery.

❗ The inguinal ligament runs from the pubic tubercle medially to the anterior superior iliac spine laterally.

Appendicitis and small bowel disease

Acute appendicitis Most common surgical emergency in the UK. *Peak age*: 10–30y. Presents with central abdominal colic that progresses to localize in the right iliac fossa. Pain is worse on movement (especially coughing, laughing) and associated with anorexia, nausea ± vomiting, dysuria, constipation, or rarely diarrhoea.

Assessment Watch for discomfort on walking (walk stooped). May be flushed and unwell—pyrexial (~37.5°C); furred tongue and/or fetor oris; tenderness, rebound tenderness and guarding in the right iliac fossa (especially over McBurney's point—⅔ of the distance between the umbilicus and anterior superior iliac spine); pain in the right iliac fossa on palpation of the left iliac fossa (Rovsing's sign). Urinalysis is normal or +ve for protein and/or leucocyte esterase but −ve for nitrites.

Differential diagnosis

- Mesenteric adenitis
- Gastroenteritis
- Meckel's diverticulum
- Intussusception
- Crohn's disease
- Urological cause, e.g. UTI, testicular torsion
- Gynaecological cause, e.g. pelvic inflammatory disease, ectopic pregnancy
- Non-abdominal cause e.g. otitis media, diabetic ketoacidosis, pneumonia

Management Admit as a surgical emergency—expect to be wrong ~½ the time. *Complications*: generalized peritonitis 2° to perforation; appendix abscess; appendix mass; subphrenic abscess; female infertility

⚠ **Appendicitis in pregnancy** Appendicitis affects 1 in 1000 pregnancies. Mortality is ↑ and perforation more common (15–20%). Fetal mortality is 5–10% for simple appendicitis; 30% when there is perforation. Due to the pregnancy, the appendix is displaced—pain is often felt in the paraumbilical region or subcostally. Admit immediately if suspected.

Children with appendicitis Symptoms/signs of appendicitis may be atypical—especially if very young—as children localize pain poorly and signs of peritonitis can be difficult to elicit.

- If unsure of diagnosis, and the child is unwell, admit
- If unsure of diagnosis, and the child is well, either arrange to review a few hours later or ask the carer to contact you if there is any deterioration or change in symptoms

Mesenteric adentitis Inflammation of the mesenteric LNs causing abdominal pain in children. May follow URTI. Can mimic appendicitis. Check MSU to exclude UTI. If guarding/rebound tenderness, refer for acute surgical assessment. Settles spontaneously with simple analgesia and fluids. If not settling in 1–2wk refer for paediatric assessment.

Subphrenic abscess Rarely follows 7–21d after generalized peritonitis—particularly after acute appendicitis. Presents with general malaise, swinging fever, nausea and weight ↓ ± pain in the upper abdomen radiating to the shoulder tip. Breathlessness can be associated due to reactive pleural effusion or lower lobe collapse. *Examination*: subcostal tenderness ± liver enlargement. FBC ↑ WCC. If suspected admit.

Meckel's diverticulum Remnant of the attachment of the small bowel to the embryological yolk sac. It is 2 inches (~5cm) long, ~2 foot (100cm) proximal to the appendix, and present in 2% of the population. A Meckel's diverticulum may not cause any problems or cause an appendicitis-like picture; acute intestinal obstruction or GI bleeding. Symptoms can occur at any age but are most common in children.

Intussusception ➜ p. 869
Crohn's disease ➜ p. 384
Coeliac disease ➜ p. 382
Obstruction and ischaemia ➜ p. 372

Adhesions Arise as a result of intra-abdominal inflammation. Bowel loops become adherent to each other, omentum, mesentery, and the abdominal wall. Fibrous bands may form connecting adjacent structures. Presents with abdominal pain ± obstruction. *Causes*: surgery; intra-abdominal sepsis (e.g. appendicitis, cholecystitis; salpingitis); inflammatory bowel disease; endometriosis. Refer to a surgeon. Treatment is difficult as any surgery may result in new adhesions; conservative management with analgesia and stool softeners is preferred. Laparoscopic, or rarely open division of adhesions, is occasionally necessary.

Intestinal non-Hodgkin's lymphoma The majority of intestinal NHLs are B-cell-type lymphomas, but coeliac disease is associated with T-cell intestinal lymphoma. *Abdominal symptoms*: non-specific abdominal pain (70–80%); perforation (up to 25%); bowel obstruction; abdominal mass; intussusception; malabsorption (usually lymphoma associated with coeliac disease) or alteration in bowel habit (small intestine NHL may present like Crohn's disease). *Systemic symptoms*: weight ↓ (30%), fatigue, sweats, unexplained fevers. ❗ Lymphadenopathy and hepatosplenomegaly are usually absent.

Management ➜ p. 656. Gastric lymphoma may remit with treatment of *H. pylori* infection.

Carcinoid tumours Slow-growing tumours of low malignancy which arise from neuroendocrine cells or their precursors. *Incidence*: 3–4/100,000. *Peak age*: 61y. ♀ > ♂. 60% are in the midgut (especially appendix and terminal ileum). Examination may reveal an abdominal mass and/or enlarged liver. Rarely presents with bowel obstruction. Ileal carcinoids are multiple in 30%. *Non-intestinal sites*: lung, testes, and ovary.

Carcinoid syndrome Affects <10% of patients with a carcinoid tumour. Develops when serotonin (5HT) is released by the tumour and not degraded by the liver due to hepatic metastases. *Features*:
- Paroxysmal flushing, e.g. following alcohol or certain foods
- Watery, explosive diarrhoea
- Abdominal pain
- Bronchoconstriction (like asthma)
- Right heart failure
- Rash—symmetrical, pruritic erythematous rash which blisters/crusts

Management Refer for urgent assessment if suspected. Therapeutic options include surgery, somatostatin analogues such as octreotide, or radiofrequency ablation of liver metastases. Prognosis—if no metastases, median survival is 5–8y; with metastases median survival is 38mo.

Colorectal cancer screening

Screening for colorectal cancer is available throughout the UK. Overall colorectal cancer 5y survival ≈50%. Patients presenting with tumour confined to the bowel wall have >90% long-term survival. Screening aims to detect colorectal cancer at an early stage to ↑ survival chances.

Screening test Faecal immunochemical test (FIT) kits are sent every 2y to all patients aged 60–74y with instructions for completion/return. Results are sent to the patients in <2wk.

Screening by a one-off flexible sigmoidoscopy is being introduced gradually across England for all patients >55y.

Screening outcomes All those who have a positive FIT test are invited for a colonoscopy pre-assessment appointment. Possible outcomes from colonoscopy are summarized in Table 12.8. If 60% of those aged 60–69y do the FIT test, 1200 deaths would be prevented each year.

Family history If a patient has one first-degree relative (mother, father, sister, brother, daughter, or son) with colorectal cancer, risk of developing colorectal cancer is ↑ 2–3×.

Refer for colonoscopy At presentation or aged 35–40y (whichever is later) and repeat colonoscopy aged 55y if:

- 2× first-degree relatives with a history of colorectal cancer *or*
- 1× first-degree relative with a history of colorectal cancer aged <45y

Refer for specialist follow-up and genetic counselling If:

- >2× first-degree relatives with a history of colorectal cancer *or*
- Family history of:
  - Familial adenomatous polyposis (FAP)—usually develop cancer aged <40y. Lifetime risk of colorectal cancer is 1:2.5
  - Juvenile polyposis—lifetime risk of colorectal cancer is 1:3
  - Peutz–Jeghers syndrome—autosomal dominant disorder. Benign intestinal (usually small intestine) polyps in association with dark freckles on lips, oral mucosa, face, palms, and soles. May cause GI obstruction or GI bleeding. Malignant change occurs in ≈3%
  - Hereditary non-polyposis colorectal cancer (Lynch syndrome)—≥3 family members with colorectal cancer where ≥2 generations have been affected and ≥1 affected family member developed the disease <50y of age; 40% lifetime risk of colorectal cancer
  - MMR (mismatch repair) oncogene

Ulcerative colitis ↑ risk of colorectal cancer. Offer all patients a follow-up plan agreed with their specialist. In some cases, prophylactic colectomy is appropriate.

Previous colorectal cancer ↑ risk of developing a second colorectal primary. After successful treatment, younger patients are routinely followed up with colonoscopy every 5y until 70y. Remain vigilant for recurrences and re-refer urgently if suspected.

Table 12.8 Colonoscopy outcomes

- ~2% of those FIT tested are referred on for colonoscopy—uptake of colonoscopy is ~80%
- Sensitivity of colonoscopy to detect significant abnormalities is ~90%
- Polyps found during colonoscopy are usually removed
- Complications of colonoscopy include heavy bleeding (1:150); bowel perforation (1:1500); death (1:10,000)

Colonoscopy result		Explanation	Action
Normal (~50%)		No abnormalities detected	FIT screening offered again in 2y if <70y
Polyp (~40%)	Low risk	1–2 small (<1cm) adenomas	FIT screening offered again in 2y if <70y
	Intermediate risk	3–4 small (<1cm) adenomas or ≥1 adenoma ≥1cm	3 yearly colonoscopy until 2× negative examinations
	High risk	≥5 adenomas or ≥3 adenomas of which at least 1 is ≥1cm	Colonoscopy at 12mo then 3 yearly colonoscopy until 2× negative examinations
Cancer (~10%)		Colorectal cancer detected at colonoscopy	Refer urgently for further treatment
Other pathology		Other pathology (e.g. UC) detected at colonoscopy	Refer/treat/advise as necessary
Technical difficulty		Unable to perform the procedure adequately	Repeat colonoscopy or alternative imaging

FIT testing for symptomatic patients FIT tests should be offered to adults without rectal bleeding who are:

- Aged ≥50y with unexplained abdominal pain or weight loss, ***or***
- Aged <60y with changes in bowel habit or iron deficiency anaemia, ***or***
- Aged ≥60y with anaemia without iron deficiency

Further information

NHS Bowel Cancer Screening Programme 🖱 https://www.gov.uk/guidance/bowel-cancer-screening-programme-overview

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

NICE (2017) Quantitative faecal immunochemical tests to guide referral for colorectal cancer in primary care. 🖱 https://www.nice.org.uk/guidance/dg30

Colorectal cancer

Lifetime risk of developing colorectal cancer is 1:15 for ♀ and 1:19 for ♂. Colorectal cancer accounts for 14% of all cancers and 16,000 deaths/y in the UK. ⅔ arise in the colon and ⅓ in the rectum; 72% of tumours occur in patients >65y and >95% are adenocarcinomas.

Adenomatous polyps Bowel cancers arise from polyps over many years. Polyps may be removed because of risk of malignant change. Follow-up surveillance with repeated colonoscopy may be necessary depending on the number of polyps and their size (Table 12.8, ➲ p. 369).

Protective and risk factors

Lifestyle factors

- Obesity—↑ risk by 15% if overweight and 30% if obese
- Dietary factors—diets with less red and processed meat, and more vegetables, fibre, fish, and milk are associated with ↓ risk (diet is thought to explain geographic variations)
- Alcohol—↑ risk for heavy drinkers—especially if also low folate
- Physical activity—↑ physical activity can ↓ risk by 30%

Medication history

- HRT—risk ↓ by 20% if ever taken; ↓ by 30% if taking HRT currently
- COC pill—risk ↓ by 18% if ever taken
- Statins—risk is ↓ after 5y use
- Aspirin—75mg od taken for >5y ↓ risk by 40%

Other medical history

- History of gallbladder disease and/or cholecystectomy—50% ↑ in risk
- Type 2 (non-insulin dependent) diabetes—30% ↑ risk
- UC or Crohn's disease—↑ risk (➲ p. 384)

Family history ➲ p.368

Bowel cancer screening ➲ p. 368

Presentation May be found at bowel cancer screening. Clinical presentation depends on site involved:

- Change in bowel habit Diarrhoea ± mucus, constipation or alternating diarrhoea and constipation, tenesmus
- Intestinal obstruction Pain, distension, absolute constipation, ± vomiting. May be an acute, sudden event (20% of patients not detected by screening present with an acute obstruction) or gradually evolve
- Rectal bleeding Bright red rectal bleeding or +ve faecal occult blood test—60% rectal tumours. Rarely melaena if high tumour
- Perforation Causing generalized peritonitis, or into an adjacent viscus (e.g. bladder) resulting in a fistula
- Spread Abdominal distension 2° to ascites, jaundice, rectal/pelvic pain
- General effects Weight ↓, anorexia, anaemia, malaise

Examination and investigation

- General examination—cachexia, jaundice, anaemia (check FBC)
- Abdominal mass
- Hepatomegaly
- Ascites
- Rectal examination—detects >75% of rectal tumours

⚠ **Suspicious lower GI symptoms and signs** Refer urgently (to be seen in <2wk) to a team specializing in lower GI malignancy^{N}:

Any age

- Rectal or abdominal mass consistent with involvement of large bowel. ❗ A pelvic mass outside the bowel warrants urgent referral to a urologist or gynaecologist
- Rectal bleeding AND abdominal pain or change in bowel habit, weight ↓ or iron deficiency anaemia

Aged ≥40y With unexplained weight ↓ and abdominal pain

Aged ≥50y With unexplained rectal bleeding

Aged ≥60y With iron deficiency anaemia or changes in bowel habit or tests showing faecal occult blood in their faeces

Specialist management Confirmation of diagnosis with sigmoidoscopy/colonoscopy and/or CT colonography. If diagnosis is confirmed further investigations include LFTs, tumour markers (carcinoembryonic antigen or CEA is produced in >80% advanced tumours), CXR, CT/MRI, and USS to evaluate spread.

Treatment Laparoscopic or open surgical resection when possible. Staging based on findings at surgery dictates further management with chemotherapy. For patients with more advanced disease, resection of or radioablation of hepatic metastases may be an option.

Adverse pathological features

- Presence/number of involved LNs
- Lymphovascular, perineural, or venous invasion
- Depth of bowel wall penetration
- Positive resection margin
- Mucinous histology

Adverse clinical features

- Emergency presentation with bowel obstruction or perforation
- Incomplete resection
- Metastatic disease
- Presentation aged <50y

Further information

NICE (2011, updated 2014) Colorectal cancer. 🖰 www.nice.org.uk/guidance/CG131

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖰 www.nice.org.uk/guidance/ng12

SIGN (2011) Diagnosis and management of colorectal cancer. 🖰 www.sign.ac.uk/sign-126-diagnosis-and-management-of-colorectal-cancer.html

Patient advice and support

British Colostomy Association ☎ 0800 328 4257 🖰 www.colostomyassociation.org.uk

Cancer Research UK ☎ 0808 800 4040 🖰 www.cancerhelp.org.uk

Macmillan Cancer Support ☎ 0808 808 0000 🖰 www.macmillan.org.uk

Other large bowel conditions

Intestinal obstruction Blockage of the bowel due to either mechanical obstruction or failure of peristalsis (ileus). *Causes*:

- Obstruction from outside the bowel Adhesions/bands; volvulus; obstructed hernia (➲ p. 364); neighbouring malignancy (e.g. bladder)
- Obstruction from within the bowel wall Tumour; infarction; congenital atresia; Hirschsprung's disease; inflammatory bowel disease (➲ p. 384); diverticulitis
- Obstruction in the lumen Impacted faeces/constipation (➲ p. 350); bolus obstruction (e.g. swallowed foreign body); gallstone ileus; intussusception (➲ p. 869); large polyps
- Ileus/functional obstruction Postoperatively; electrolyte disturbance; uraemia; DM; back pain; anticholinergic drugs

Presentation Anorexia; nausea; vomiting (may be feculent) gives relief; colicky central abdominal pain and distension; absolute constipation for stool and gas (though if high obstruction constipation may not be absolute). *Examination*: uncomfortable and restless; abdominal distention ± tenderness (though no guarding/rebound); active tinkling bowel sounds or quiet/absent bowel sounds (later).

Management Admit as surgical emergency.

Diverticulosis Common condition of the colon associated with muscle hypertrophy and ↑ intraluminal pressure. Mucosa-lined pouches are pushed out through the colonic wall usually at the entry points of vessels. These pouches are the diverticula; 95% are in the sigmoid colon although they may occur anywhere in the bowel. They are present in >1:3 people >60y in the UK. Risk factors include low-roughage diet and age. Diverticular disease implies the diverticula are symptomatic—Table 12.9.

Ischaemic bowel Interruption of the blood supply of the bowel.

- 1° ischaemia Usually due to either mesenteric embolus from the right side of the heart, or venous thrombosis and typically occurs in elderly patients who might have pre-existing heart or vascular disease
- 2° ischaemia Usually due to intestinal obstruction (e.g. strangulated hernia, volvulus, intussusception)

Presentation Sudden onset of abdominal pain which rapidly becomes severe. There may be a prior history of pain worse after meals (mesenteric angina). Rarely presents with PR bleeding. *Examination*: very unwell; shocked; may be in AF; generalized tenderness but normally no guarding/rebound. Often signs are out of proportion to symptoms.

Management Give opioid analgesia. Admit as surgical emergency.

Sigmoid volvulus Occurs in people who have redundant colon on a long mesentery with a narrow base. The sigmoid loop twists causing intestinal obstruction. The loop may become ischaemic. *Risk factors*: constipation, laxatives, tranquillizers. Presents with acute onset of abdominal distention and colicky abdominal pain with complete constipation and absence of flatus. There may be a history of repeated attacks.

Management Admit acutely to hospital. Treatment is release by passing a flatus tube and/or surgery. Once treated, ↓ recurrences by preventing constipation and stopping tranquillizers if possible.

Table 12.9 Presentation and management of diverticular disease

	Presentation	Management
Chronic diverticulitis (painful diverticular disease)	Presents with altered bowel habit, abdominal pain (often colicky and left sided), nausea, and flatulence Symptoms are often improved by defecation	Investigate for change in bowel habit (➔ p. 370) Once diverticular disease is confirmed, treat with high-fibre diet ± antispasmodics (e.g. mebeverine 135mg tds) Refer if severe symptoms
Acute diverticulitis	Presents with: • Altered bowel habit • Colicky left-sided abdominal pain—may become continuous and cause guarding/peritonism in the left iliac fossa • Fever • Malaise ± nausea • Flatulence ❗ There may be few abdominal signs in the elderly	Treat with oral antibiotics (e.g. co-amoxiclav 250mg tds or cefaclor 250–500mg tds and metronidazole 400mg bd or ciprofloxacin 500–750mg bd) There may also be some benefit from a low-residue diet If severe symptoms, uncertain diagnosis, or not settling, admit as an acute surgical emergency
Diverticular abscess	Presents with swinging fever, general malaise, ± other localizing symptoms, e.g. pelvic pain	Refer for urgent surgical assessment/admit as a surgical emergency
Perforated diverticulum	Presents with ileus, peritonitis and shock	Admit as an acute surgical emergency
Fistula formation	A fistula may form if a diverticulum perforates into bladder, vagina, or small bowel—➔ p. 352	Refer for surgical assessment Treatment is usually surgical
Diverticular haemorrhage	Common cause of rectal bleeding— usually sudden and painless	Gain IV access Admit as an acute surgical emergency (➔ p. 1061)
Post-infective stricture	Fibrous tissue formation following infection can cause narrowing of the colon → obstruction	Keep stool soft If recurrent problems refer for surgery

Hirschsprung's disease Caused by absence of the ganglion cells of the myenteric plexus in the distal bowel. Presents with delay in passing meconium, abdominal distension, vomiting, and poor feeding in a neonate. If only a short segment is affected, presentation may be much later with chronic constipation. Diagnosis is confirmed with rectal biopsy. Refer to surgery. Treatment is surgical removal of the affected area of bowel.

Carcinoma of the colon ➔ p. 370

Anal conditions ➔ p. 374

Inflammatory bowel disease ➔ p. 384

Anal and perianal problems

Haemorrhoids ('piles') Common in all age groups from mid-teens onwards. Represent distention of the submucosal plexus of veins in the anus. 3 main groups situated at 3, 7, and 11 o'clock positions (relative to the patient viewed in lithotomy position). *Risk factors*: constipation; FH; varicose veins; pregnancy; ↑ anal tone (cause not understood); pelvic tumour; portal hypertension. *Classification*:

- **1st degree** Piles remain within the anal canal
- **2nd degree** Prolapse out of anal verge but spontaneously reduce
- **3rd degree** Prolapse out of anus and require digital reduction
- **4th degree** Permanently prolapsed

Presentation Discomfort or discharge ± fresh red rectal bleeding (blood on toilet paper, coating stool, or dripping into pan after defecation); feeling of incomplete emptying of the rectum; mucus discharge; pruritus ani. *Rectal examination*: prolapsing piles are obvious, 1st-degree piles are not visible or palpable.

Management If piles are not obvious on examination, arrange proctoscopy ± sigmoidoscopy for all patients >40y. *Treatment*: soften stool (bran, ispaghula husk) and recommend topical analgesia (e.g. lidocaine 5% ointment or OTC preparation). If not responding to treatment, uncertainty over diagnosis, or severe symptoms (e.g. soiling of underwear), refer for surgical assessment. *Complications*:

- **Strangulation** Circulation to the pile is obstructed by the anal sphincter. Results in intense pain + anal sphincter spasm. Treat with analgesia. If severe pain or symptoms are not settling, admit
- **Thrombosis** Pain/anal sphincter spasm—analgesia, ice packs and bed rest—consider referral for surgery to prevent recurrence

Perianal haematoma (thrombosed external pile) Due to a ruptured superficial perianal vein causing a subcutaneous haematoma. Presents with sudden onset of severe perianal pain. A tender, 2–4mm 'dark blueberry' under the skin adjacent to the anus is visible. Give analgesia. Settles spontaneously over ~1wk. If <1d old can be evacuated via a small incision under LA.

Rectal prolapse Occurs in 2 age groups—the very young, and those >60y. Presents with mass coming down through the anus ± anal discharge. In adults there are 2 types:

- **Mucosal** Adults with 3rd-degree piles—bowel musculature remains in position but redundant mucosa prolapses from the anal canal
- **Complete** Descent of the upper rectum into the lower anal canal. Usually due to weak pelvic floor from childbirth. Bowel wall is inverted and passed out through the anus. May be associated uterine prolapse

Refer for surgery. A supporting ring may be used if unfit for surgery.

Anal fissure Anal mucosa is torn—usually on the posterior aspect of the anal canal. May occur at any age. Presents with pain on defecation ± constipation ± fresh rectal bleeding ('blood on toilet paper'). The fissure is often visible as is a 'sentinel pile' (bunched up mucosa at the base of the tear). Rectal examination is very tender due to muscle spasm.

Management Soften stool (e.g. ispaghula husk); try analgesic suppositories/cream (e.g. cinchocaine/hydrocortisone). If unsuccessful add glyceryl trinitrate 0.4% ointment bd which relieves pain and spasm but may cause headache; 2% topical diltiazem cream bd is a 3rd-line option (unlicensed). If interventions fail refer for surgical review.

Perianal abscess Usually caused by infection arising in a perianal gland. Tends to lie between the internal and external sphincters and points towards the skin at the anal margin. May affect patients of any age and presents with gradual onset of perianal pain which becomes throbbing and severe; defecation and sitting are painful—characteristically patients sit with one buttock raised off the chair. *Examination*: abscess in the skin next to the anus. Refer as an acute surgical emergency for drainage.

Perianal fistula Abnormal connection between the lumen of the anus (or rectum) and skin. Usually develops from a perianal abscess. Fistulae are either 'high' (open into the bowel above the deep external anal sphincter) or 'low' (open into the bowel below this point). High fistulae are rare and usually due to UC, Crohn's disease, or tumour—they are more complex to repair. Presents with persistent perianal discharge and/or recurrent abscess. The external opening is usually visible lateral to the anus; the internal opening may be palpable on rectal examination. Refer for surgical repair.

Pilonidal sinus Obstruction of a hair follicle in the natal cleft. The ingrowing hair triggers a foreign body reaction → pain, swelling, abscess, and/or fistula formation ± foul smelling discharge. Refer for surgery.

Pruritus ani Itching around the anus. Occurs if the anus is moist or soiled, e.g. poor personal hygiene; anal leakage or faecal incontinence; fissures; nylon/tight underwear. *Other causes*: dermatological conditions (e.g. contact dermatitis, lichen sclerosus); threadworm infection; anxiety; other causes of generalized pruritus (➲ p. 566). Treat cause if possible; avoid spicy food; moist wipe post-defecation.

Threadworm Common in the UK—especially in children. *Enterobius vermicularis* causes anal itch as it leaves the bowel to lay eggs on the perineum. Often seen as silvery thread-like worms at the anus of children. *Treatment*: mebendazole (available OTC). Treat household contacts as well as the index case.

Anal ulcers Rare. Consider Crohn's disease, syphilis, tumour—refer[N].

Anal cancer Usually squamous cell cancer (>50%). *Risk factors*: anal sex; syphilis; anal warts (HPV). Presents with bleeding, pain, anal mass or ulcer, pruritus, stricture, change in bowel habit. A mass may be palpable on rectal examination. Check for inguinal LNs.

Management Refer for urgent surgical review and confirmation of diagnosis. Treatment is usually with a combination of radiotherapy ± chemotherapy. Abdominoperineal resection is reserved for salvage therapy after chemo or radiotherapy failure.

Patients with ostomies

❗ Specialist stoma nurses are an extremely useful source of advice and help. If in doubt about the correct stoma appliances and accessories to supply, or a patient has a problem with a stoma, wherever possible liaise with your local specialist stoma nurse.

The first iatrogenic stoma was constructed in France in 1776 for an obstructing rectal cancer. Stomas (from the Greek meaning 'mouth') may be temporary or permanent (Table 12.10).

Stoma retraction Can lead to leakage and severe skin problems. Most common reason for re-operation. Refer for specialist advice.

Prolapse Seen most frequently with loop colostomy. If persists and disrupts pouching, refer for consideration of revision.

Peristomal hernia Common complication. Symptomatic cases require referral for repair.

Stenosis Narrowing of the stoma may result in difficulty or pain passing stool and/or obstruction. If problematic refer for revision.

Skin complication Skin irritation can be due to:

- Leakage onto the skin
- Allergic reactions to the adhesive material in a skin barrier
- Fungal infection
- Inadequate hygiene

Prevention of skin complications

- Advise patients to clean, rinse, and pat the skin dry between pouch changes
- Avoid using an oily soap, which can leave a film that interferes with proper adhesion of the skin barrier
- Ensure the pouch system fits
- Treat any infection with oral antibiotics and/or oral/topical antifungals
- Apply skin barrier cream
- If the skin is uneven (e.g. due to scarring), fill irregularities with stoma paste to give a better fit
- Consider the use of convex discs or stoma belts (refer to specialist stoma nurse for advice)

Diet

- Avoid foods that cause intestinal upset or diarrhoea
- For descending/sigmoid colostomy, avoid foods that cause constipation. If constipation does occur, ↑ fluid intake and/or dietary fibre
- Certain foods, e.g. beans, cucumbers, and carbonated drinks, can cause gas, along with certain habits such as talking or swallowing air while eating, using a straw, breathing through the mouth, and chewing gum
- A daily portion of applesauce, cranberry juice, yogurt, or buttermilk can help control odour. If odour is strong and persistent, consider use of charcoal filters or pouch deodorizers (seek advice from a specialist stoma nurse)

Table 12.10 The 3 main types of stoma

Colostomy	Ileostomy	Urostomy
Age: most >50y	*Peak age range:* 10–50y	*Age:* most >50y
Output: depends on site: • Transverse colostomy—soft stool • Descending/sigmoid colostomy—formed stool	*Output:* soft/fluid stool	*Output:* urine—continent procedures using bowel to fashion a bladder which is then drained with a catheter through the stoma are becoming common
Reasons for colostomy: Carcinoma Diverticular disease Trauma Radiation enteritis Bowel ischaemia Hirschsprung's disease Congenital abnormalities Obstruction Crohn's disease Faecal incontinence	*Reasons for ileostomy*: Ulcerative colitis Crohn's disease Familial polyposis coli Obstruction Radiation enteritis Trauma Bowel ischaemia Meconium ileus Carcinoma	*Reasons for urostomy*: Carcinoma Urinary incontinence Fistulas Spinal column disorders

Drugs Enteric-coated and modified-release preparations are unsuitable for people with bowel stomas—particularly for patients with ileostomy.

Psycho-social problems Self-help groups provide information and tips on lifestyle and stoma care; specialist stoma nurses can provide support and counselling.

Activities Advise patients to avoid rough contact sports and heavy lifting as these might → herniation around the stoma. Patients with stomas may swim. Water will not enter a stoma due to peristalsis so stomas do not need to be covered when bathing. A body belt (available on FP10) to hold the stoma bag in place against the body may stop rustling/leakage for those doing aerobic exercise—seek advice from a specialist stoma nurse.

Travel Advise patients to pack sufficient supplies of their stoma products and carry supplies with them in case baggage is misplaced. Avoid storing supplies in a very hot environment as heat may damage pouches.

Patient advice and support

British Colostomy Association ☎ 0800 328 4257 🖱 www.colostomyassociation.org.uk

Chronic diarrhoea and malabsorption

Chronic diarrhoea Diarrhoea persisting >4wk. Patients' perceptions of diarrhoea vary widely. Clarify what is meant. Chronic diarrhoea affects ~4–5% of adults in the UK. There are many causes (Table 12.11) and all patients require investigation. Careful history is vital.

Symptoms suggestive of organic disease

- History of <3mo duration
- Mainly nocturnal or continuous (as opposed to intermittent) diarrhoea
- Significant weight ↓
- Liquid stools with blood and/or mucus

Symptoms suggestive of malabsorption

- Pale and/or offensive stools
- Steatorrhoea—excess fat in faeces. The stool is pale-coloured, foul smelling, and floats ('difficult to flush')

Examination and investigation Full examination. Look for signs of systemic disease and examine abdomen/pelvis thoroughly. Check:

- **Blood** FBC, ESR, Ca^{2+}, LFTs, haematinics, TFTs, coeliac serology, CA125 in women (refer for USS of abdomen/pelvis if ↑)
- **Stool** M,C&S ± faecal calprotectin (useful in primary care for distinguishing between IBS and inflammatory bowel disease if <40y)

Management

- If obvious identifiable cause (e.g. GI infection, constipation, drug side effect) treat and review. Refer to gastroenterology if treatment fails
- If symptoms suggestive of functional bowel disease and <45y with normal investigations, irritable bowel syndrome is likely. Reassure, offer advice, and review as necessary. Refer to gastroenterology if atypical symptoms appear or the patient is unhappy with the diagnosis
- Otherwise refer to gastroenterology for assessment. Speed of referral depends on age and severity of symptoms

⚠ **Refer urgently** (to be seen in <2wk)[N]

To a team specializing in colorectal cancer if

- Any age + anal, rectal, or abdominal mass*
- Any age + rectal bleeding + unexplained abdominal pain, change in bowel habit, weight loss, or iron deficiency anaemia
- Aged ≥40y + unexplained weight loss + abdominal pain, or
- Aged ≥50y + unexplained rectal bleeding, or
- Aged ≥60y + iron-deficiency anaemia or persistent change in bowel habit or faecal occult blood +ve

* A pelvic mass outside the bowel warrants urgent referral to a urologist or gynaecologist.

For direct access CT (or USS if CT not available) if ≥60y + weight loss AND ≥1 of:

- Diarrhoea
- Back pain
- Abdominal pain
- Nausea/vomiting
- Constipation
- New-onset diabetes

Table 12.11 Causes of chronic diarrhoea

Colon
Colonic cancer
Ulcerative colitis
Crohn's disease
Constipation with overflow diarrhoea

Endocrine
DM (autonomic neuropathy)
Hyperthyroidism
Hypoparathyroidism
Addison's disease
Hormone-secreting tumours (e.g. carcinoid)

Small bowel
Crohn's disease
Coeliac disease
Other enteropathies (e.g. Whipple's disease)
Bile acid malabsorption
Ischaemia
Enzyme deficiencies (e.g. lactase deficiency)
Radiation damage
Bacterial overgrowth
Lymphoma
Infection (e.g. giardiasis, *Cryptosporidium*)
Irritable bowel syndrome

Pancreas
Pancreatic cancer
Chronic pancreatitis
CF

Other
Ovarian cancer
Bowel resection
Bile salt malabsorption
Intestinal fistula
Drugs
Alcohol
Autonomic neuropathy
'Factitious' diarrhoea

Malabsorption Presents with chronic diarrhoea, weight ↓, steatorrhoea, vitamin/iron deficiencies, and/or oedema due to protein deficiency. Refer to gastroenterology for investigation/treatment of the cause.

Usual causes

- Coeliac disease—➔ p. 382
- Chronic pancreatitis—➔ p. 402
- Crohn's disease—➔ p. 384

Rarer causes

- Cystic fibrosis (CF)
- Pancreatic cancer—➔ p. 404
- Whipple's disease
- Biliary insufficiency
- Bacterial overgrowth
- Chronic infection (e.g. giardiasis, tropical sprue)
- Following gastric surgery

Whipple's disease A cause of malabsorption which usually occurs in ♂ >50y. *Other features*: arthralgia, pigmentation, weight ↓, lymphadenopathy, ± cerebellar or cardiac signs. *Cause: Tropheryma whipplei*. Refer for gastroenterology assessment. Jejunal biopsy is characteristic. *Treatment*: long-term broad spectrum antibiotics.

Malabsorption in children ➔ p. 866

Factitious diarrhoea Responsible for 4% of referrals to gastroenterology departments and 20% of tertiary referrals. Due to laxative abuse or adding of water or urine to stool samples. Difficult to spot—have a high index of suspicion especially in patients with history of eating disorder or somatization.

Further information

British Society of Gastroenterology (2018) Guidelines for the investigation of chronic diarrhoea. 🖱 https://gut.bmj.com/content/67/8/1380
NICE (2009) Diarrhoea and vomiting in children under 5. 🖱 www.nice.org.uk/Guidance/CG84
NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Faecal incontinence

Affects ~2% of all ages, causing great personal disability. It is a common reason for carers to request placement in a nursing home.

Causes

- Age and frailty
- Constipation (overflow incontinence)
- Childbirth
- Colonic resection/anal surgery
- Rectal prolapse/haemorrhoids
- Loose stools or diarrhoea from any cause, e.g. inflammatory bowel disease
- After radiotherapy
- Systemic sclerosis
- Neurological disorders
- Cognitive deficit
- Congenital disorders (e.g. anal atresia, Hirschsprung's disease)
- Emotional problems (e.g. encopresis in children)

History Aimed at establishing the underlying causes of the incontinence (may be >1) and other factors that might be contributing to it. *Ask about*:

- Onset and nature of symptoms. ❗ Always consider faecal incontinence when patients present with anal soreness and/or itching
- Bowel habit including timing and frequency of incontinence
- Difficulties with toileting and help available
- Other medical conditions
- Medication
- Diet
- Social circumstances

Examination General and rectal examination (to detect abnormalities of anal tone, local anal pathology, e.g. rectal prolapse, and constipation causing overflow incontinence). Further examination depends on age group and history, e.g. cognitive assessment if suspected cognitive deficit; neurological examination if ↓ anal tone.

⚠ Persistent change in bowel habit to looser stools may be a sign of GI malignancy—➜ p. 362.

Primary care management

Treatment of cause

- Clear any constipation/faecal loading (➜ p. 350)—use rectal preparations initially to clear faecal load. If unsuccessful/rectal preparations are inappropriate, then switch to oral laxatives. Take steps to prevent recurrence, e.g. add fibre to diet, ↑ fluid intake, consider regular laxatives
- Treat other reversible causes, e.g. infective diarrhoea, UC
- Consider alternatives to any contributing medications, e.g. tranquillizers

General measures where cause cannot be treated

- Advise fluid intake of at least 1.5L/d
- Encourage bowel emptying after a meal—advise patients to assume a seated/squatting position and not to strain
- Ensure that toilet facilities are private, accessible, and safe—refer for OT assessment if needed

- Manipulate diet to promote optimal stool consistency and predictable bowel emptying. A food/fluid diary may be helpful. Only change one food at a time. Consider referral to a dietician
- If stool must be in the rectum at a set time (e.g. when a carer is there), manipulate bowel action with PR/PO laxatives and/or loperamide
- If loose stools, consider treatment with loperamide, co-phenotrope or codeine phosphate, prn or continuously. When using loperamide, introduce at a very low dose (consider syrup for doses <2mg) and ↑ dose until desired stool consistency is reached. Dose and/or frequency can be adjusted ↑ or ↓ in response to stool consistency and lifestyle. ❗ Do not use if hard stools, undiagnosed diarrhoea, or flare-up of UC
- Review regularly. If no improvement with simple strategies, consider referral for specialist care

Patients with faecal incontinence from enteral feeding Discuss with the patient's dietician. Modifying type/timing of feeds may help.

Patients with spinal injury or disease Bowel function is a reflex action which we learn to override as children. If the lesion is above the level of this reflex pathway (T12 for bowel function) then automatic emptying will still occur when the bladder or bowel is full, although there is no control. If the lesion is below this level there is no emptying reflex. Bowel care programmes reflect this. Useful leaflets are available from the Spinal Injuries Association (☎ 0800 980 0501 🖱 www.spinal.co.uk).

Referral Consider if symptoms are not controlled:

- To continence adviser—for advice on skin care/hygiene, and supplies of incontinence pads. Pelvic floor muscle training, bowel retraining, biofeedback, electrical stimulation, and/or rectal irrigation may be useful. Devices, e.g. anal plugs or faecal collectors, can help in some situations
- To surgeon—for sphincter repair if significant sphincter defect; for consideration of implanted sacral nerve stimulation device; for appendicostomy/continent colonic conduit for anterograde irrigation in patients with colonic motility disorders; for stoma formation (last resort)
- To old age psychiatry—if cognitive deficit and incontinence
- To paediatrics—if encopresis due to chronic constipation—or child psychiatry if encopresis due to emotional distress

Encopresis in children ➔ p. 893

Further information

NICE (2007) Faecal incontinence. 🖱 www.nice.org.uk/guidance/cg49

Patient information

Bladder and Bowel Community Provides information and support as well as 'Just can't wait' or JCW cards. This card allows patients with bowel problems access to staff toilet facilities in many high street stores on production of their access card. ☎ 0800 031 5412 🖱 www.bladderandbowel.org

RADAR keys The National Key Scheme (NKS) offers independent access to disabled people to around 7000 locked public toilets around the UK. Keys are available to purchase from 🖱 www.disabilityrightsuk.org. If the patient has an ongoing disability, purchase can be made VAT free.

Coeliac disease

Coeliac disease is a common disorder (UK prevalence 0.5–1%, ♀:♂ ≈3:1) although only a minority have recognized disease. Gluten sensitivity results in inflammation of the bowel and malabsorption. Peak incidence in adults is in the 5th decade; in children at ~4y. Associated with HLA-DQ2 or DQ8; first-degree relatives have a 1:10 chance of being affected. See Table 12.12.

Investigation

- **Serological testing** IgA anti-tissue transglutaminase antibodies (TTG) or anti-endomysial antibodies (EMA). Test if[N]: persistent/unexplained GI symptoms, faltering growth, prolonged fatigue, unexpected weight ↓, severe/persistent mouth ulcers, unexplained iron, vitamin B$_{12}$ or folate deficiency, type 1 DM or autoimmune thyroid disease (at diagnosis), irritable bowel syndrome, first-degree relative with coeliac disease,
  ❗ test only if eaten >1 meal/d containing gluten for ≥6wk
- **Other tests** Also consider FBC, ESR/CRP, vitamin B$_{12}$, folate, ferritin, LFTs, Ca^{2+}, TFTs, and stool sample for M,C&S (if diarrhoea)

> ⚠ IgA deficiency is ↑ >6× in people with coeliac disease. If IgA deficient, IgA TTG/EMA may give false −ve result. If clinical suspicion and IgA TTG/EMA is −ve, check serum IgA. If deficient, request IgG TTG/EMA.

Initial management Refer for specialist review if:

- +ve serology—duodenal biopsy showing villous atrophy is diagnostic
- Strong clinical suspicion of coeliac disease but −ve serology
- Unwilling to reintroduce gluten to diet to enable serological testing

Gluten-free diet Cornerstone of management of coeliac disease. Should be followed lifelong. Avoid proteins derived from wheat, rye or barley. 💣 Avoidance of oats is controversial. Refer to a dietician for specialist advice. Coeliac UK provides a directory of approved products as well as recipes for those on gluten-free diets.

Prescriptions for gluten-free foods Prescribe adequate gluten-free foods (Table 12.13), marking prescriptions 'ACBS'. Add deficient nutrients (e.g. iron, folic acid, Ca^{2+}) until established on a gluten-free diet.

Failure to respond to diet Most commonly due to continued gluten ingestion (intentional or inadvertent). Re-refer to dietician. If symptoms recur after a period of remission, re-refer for specialist review.

Pneumococcal vaccination Pneumococcal infection is more common 2° to hyposplenism—advise vaccination.

Follow-up Every 12mo by GP. Routine checks include: symptoms, weight, diet, and consider blood tests (Hb, vitamin B$_{12}$, folate, iron, albumin, Ca^{2+}, TTG or EMA antibodies), co-morbidities, and need for specialist follow-up.

Long-term complications Almost eliminated by strict diet:

- Osteoporosis—consider DEXA scan at diagnosis, after 3y on a gluten-free diet (if abnormal baseline DEXA), at the menopause for ♀, aged 55y for ♂, or if fragility fracture[G]
- Malignancy—lymphoma or carcinoma of the small intestine. Rare—if suspected, refer urgently for specialist review

Table 12.12 Presentation of coeliac disease

Symptoms and signs	Associated conditions	
Chronic/intermittent diarrhoea (50%) Failure to thrive/faltering growth in children Recurrent abdominal pain/cramping/bloating Other persistent unexplained GI symptoms, e.g. nausea/ vomiting Sudden or unexpected weight ↓ Unexplained anaemia (iron deficiency or other) *Genetic predisposition* First-degree relative (parent, sibling, child) Down's/Turner syndrome	*GI* Dental enamel defects Mouth ulcers Irritable bowel syndrome Microscopic colitis Persistent/unexplained constipation Unexplained, persistent ↑ in liver enzymes (usually normalize in <6mo on gluten-free diet) Autoimmune liver disease *Musculoskeletal* ↓ bone mineral density Low trauma fracture Metabolic bone disease (e.g. rickets, osteomalacia) Sjögren's syndrome Sarcoidosis	*Endocrine* Type 1 DM Autoimmune thyroid disease Addison's disease Amenorrhoea *Other* Unexplained alopecia Dermatitis herpetiformis Depression or bipolar disorder Polyneuropathy Epilepsy Autoimmune myocarditis Chronic TTP Lymphoma Recurrent miscarriage Unexplained subfertility

Table 12.13 Guide to the amount of gluten-free products to prescribe monthly for patients with coeliac disease

Child age	Units/mo	♂ age	Units/mo	♀ age	Units/mo
1–3y	10	19–59y	18	19–74y	14
4–6y	11	60–74y	16	75+y	12
7–10y	13	75+y	14	Breastfeeding	Add 4 units
11–14y	15			3rd trimester pregnancy	Add 1 unit
15–18y	18	High activity level (♂ or ♀)—add 4 units			

400g of bread or rolls or baguette = 1 unit

250g of pasta = 1 unit

500g of bread or flour = 2 units

2 pizza bases = 1 unit

200g of sweet or savoury biscuits, crackers, or crispbread = 1 unit

Further information

British Society of Gastroenterology (2014) Diagnosis and management of adult coeliac disease. 🖱 www.bsg.org.uk/clinical-guidelines/small-bowel-nutrition/guidelines-on-the-diagnosis-and-management-of-adult-coeliac-disease.html

NICE (2015) Coeliac disease: recognition, assessment and management. 🖱 www.nice.org.uk/guidance/ng20

Patient advice and support

Coeliac UK ☎ 0333 332 2033 🖱 www.coeliac.org.uk

Inflammatory bowel disease

Ulcerative colitis (UC) and Crohn's disease are collectively termed inflammatory bowel disease.

Both are chronic, relapsing–remitting diseases characterized by acute, non-infectious inflammation of the gut. In UC, inflammation is limited to the colorectal mucosa. Extent varies from disease limited to the rectum (proctitis) to disease affecting the whole colon (pancolitis). In Crohn's, any part of the gut from mouth to anus can be affected with normal bowel between affected areas (*skip lesions*).

Cause Unknown. Both diseases are thought to result from an environmental trigger on genetically susceptible individuals. Factors implicated (none proven) include:

- Smoking—protective against UC (95% are non-smokers or ex-smokers) but a causative factor in Crohn's disease (⅔ are smokers and smoking cessation halves the relapse rate)
- Gut flora or other infections, e.g. *Mycobacterium paratuberculosis*
- Food constituents

Features Table 12.15

Differential diagnosis

- Irritable bowel syndrome
- Coeliac disease
- Anal fissure
- Gut infection, e.g. giardiasis
- Diverticulitis
- Colonic tumour
- Food sensitive colitis (infants)
- Pseudomembranous colitis
- Ischaemic colitis
- Microscopic colitis

Suspected diagnosis ~50% of severe attacks of UC are first attacks in patients who do not have a prior diagnosis. If bloody diarrhoea + fever >37.5°C or tachycardia >90bpm, admit as an acute emergency. If persistent, unexplained diarrhoea lasting >4wk and/or persistent abdominal pain, refer for urgent further investigation to exclude GI malignancy and establish diagnosis.

Assessing severity Table 12.14

Table 12.14 Assessing the severity of ulcerative colitis

Severity	Symptoms	Action
Mild	<4 liquid stools/d Little/no rectal bleeding No signs of systemic disturbance	Manage in primary care
Moderate	4–6 liquid stools/d Moderate rectal bleeding Some signs of systemic disturbance Mild disease that does not respond to treatment	Consider admission; contact specialist team for management advice
Severe	>6 liquid stools/d Severe rectal bleeding Any systemic disturbance (↑ pulse rate >90bpm, pyrexia >37.5°C, ↑ ESR, ↑ WCC, ↓ Hb <10 g/dL) Signs of malnutrition (e.g. albumin <35g/dL) Weight loss >10%	Admit as an emergency

Table 12.15 Features of inflammatory bowel disease

	UC	Crohn's disease
Incidence	10–20/100,000/y	5–10/100,000/y and increasing
Prevalence	100–200/100,000	50–100/100,000
Peak age	40–60y (85% <60y)	
Gender	♂ = ♀	
Risk factors	Smoking is protective	Smoking is a risk factor
GI symptoms	Diarrhoea + blood/mucus (stool may be solid if rectal disease only) Faecal urgency/incontinence Tenesmus Lower abdominal pain	Diarrhoea ± blood/mucus Malabsorption Abdominal pain (crampy) Mouth ulcers Bowel obstruction due to strictures Fistulae (often perianal) Abscesses (perianal and intra-abdominal)
Systemic symptoms	Tiredness and/or malaise Weight ↓ or failure to thrive/grow (children) Fever	
Associated conditions	*Joint disease*—arthritis, sacroiliitis, ankylosing spondylitis *Eye disease*—iritis or uveitis *Skin changes*—erythema nodosum, pyoderma gangrenosum (UC > Crohn's) *Liver disease*—autoimmune hepatitis (UC), gallstones (Crohn's), sclerosing cholangitis (UC > Crohn's) *Miscellaneous*—thromboembolism, osteoporosis (Crohn's), amyloidosis (Crohn's)	
Examination	*Abdominal + rectal examination*—abdominal tenderness. Anal and perianal lesions (pendulous skin tags, abscesses, fistulae) and/or mass in the right iliac fossa are characteristic of Crohn's disease *General examination*—clubbing, aphthous ulcers in the mouth (Crohn's), signs of weight loss, anaemia or hypoproteinaemia	
Investigation	*Blood*—FBC (anaemia, ↑ WCC), ESR (↑ when disease is active), eGFR, LFTs (including serum albumin). In severe UC, CRP >45g/dL after 3d steroid treatment indicates high (~85%) risk for colectomy *Stool*—M,C&S (including *Cl. difficile*) to exclude infection *AXR*—consider to clarify extent of disease, exclude toxic megacolon (transverse colon diameter >5cm) or bowel obstruction and/or identify proximal constipation. *Proctoscopy*—inflammation and shallow ulceration extending proximally from the anal margin suggests UC	

UC and Crohn's disease are rare in childhood. Presentation is variable and can be with non-specific features (e.g. failure to thrive), GI symptoms (e.g. malabsorption, bloody diarrhoea, acute abdomen), or complications (e.g. arthropathy or iritis). If suspected refer for confirmation of diagnosis and specialist management.

Management of ulcerative colitis

Active disease

- Mesalazine 2–4g daily. Topical 5-ASA derivatives are a useful adjunct if troublesome rectal symptoms
- Add steroids (prednisolone 40mg od po + rectal preparation) if prompt response is needed or mesalazine is unsuccessful. Review frequently and ↓ dose over 8wk. Rapid withdrawal ↑ risk of relapse
- Azathioprine is added if the patient is having recurring attacks despite mesalazine maintenance, frequent steroids (≥2 courses/y), disease relapses as dose of steroid is ↓, or relapse <6wk after stopping steroids. Requires regular supervision
- Ciclosporin or infliximab (anti-TNF antibody)—consultant supervised—may be effective as acute therapy for severe, steroid-refractory disease

⚠ *Admit acutely if*

- Severe abdominal pain (especially if associated with tenderness)
- Severe diarrhoea (>8×/d) ± bleeding
- Dramatic weight loss
- Fever >37.5°C, tachycardia >90bpm or other signs of systemic disease

Maintenance treatment Follow-up in 2° care is routine. Most patients require lifelong therapy. Mainstays of treatment are 5-ASA derivatives (e.g. mesalazine 1–2g/d or balsalazide 2.5g/d). Use a rectal formulation (e.g. mesalazine 1g/d PR) if disease is confined to the rectum or descending colon. Long-term treatment ↓ risk of colonic cancer by 75%. 10% are intolerant to 5-ASA derivatives—alternative is azathioprine (see earlier in topic). Treat proximal constipation with stool bulking agents or laxatives. NSAIDs can precipitate relapse so avoid.

Surgery Last resort—but should not be delayed if severe colitis and failing to respond to medical therapy. 20–30% of patients with pancolitis require colectomy—1 in 3 develop pouchitis (non-specific inflammation of the ileal reservoir) within 5y of surgery.

Prognosis At any time 50% are asymptomatic, 30% have mild symptoms, and 20% moderate/severe symptoms. <5% are free from relapse after 10y. Relapses usually affect the same part of the colon.

Complications Table 12.17

Management of Crohn's disease

Active ileal and/or colonic disease

- Treat with mesalazine 4g daily. Less effective than for UC
- Add steroids (prednisolone 40mg od po or budesonide 9mg daily) if unresponsive to mesalazine. Review frequently and ↓ dose over 8wk. Rapid withdrawal ↑ risk of relapse. ❗ steroids are associated with ↑ risk of severe sepsis and mortality in Crohn's disease so alternatives to steroid therapy are increasingly sought and steroid maintenance for >3mo should always be avoided
- Elemental or polymeric diets for 4–6wk can be a useful adjunct or alternative to steroid treatment—take consultant advice
- Other treatments (consultant supervision) include metronidazole, azathioprine, antitumour necrosis factor (infliximab or adalimumab)

- Surgery is an option if medical treatment has failed. 50% need surgery <10y after onset. Surgery is not curative and 50% will require a further operation at a later stage. After ileal resection check vitamin B_{12} levels annually

⚠ *Admit acutely if*
- Severe abdominal pain (especially if associated with tenderness)
- Severe diarrhoea (>8×/d) ± bleeding
- Dramatic weight loss
- Bowel obstruction
- Fever/other signs of systemic disease

❗ For disease elsewhere, take specialist advice.

Maintenance treatment Follow-up in 2° care is routine. Treatment is aimed at ↓ impact of the disease. Mesalazine has limited benefit. It is ineffective at doses <2g/d. Other agents used include azathioprine, mercaptopurine, methotrexate, and infliximab/adalimumab. All require consultant supervision. Treat diarrhoea symptomatically with codeine phosphate or loperamide unless it is due to active colonic disease. Colestyramine (4g 1–3 ×/d) ↓ diarrhoea due to terminal ileal disease/resection. NSAIDs can precipitate relapse—so avoid.

Prognosis 75% are back to work after the first year but 15% remain unable to work long term. Complications—Table 12.16.

Table 12.16 Complications of inflammatory bowel disease

UC	Crohn's
Toxic megacolon—colon distends and may perforate *Colonic cancer*—risk ↑ if disease >8y, onset in childhood/adolescence, age >45y, FH of colon cancer, extensive colitis, sclerosing cholangitis. Prevention: screening with colonoscopy. Frequency depends on severity of the disease and duration of symptoms *Sclerosing cholangitis*—fibrosis and stricture of intra- and extra-hepatic bile ducts. Presents with obstructive jaundice	*Intra-abdominal abscess* *Intestinal stricture*—common—may require surgery *Toxic megacolon*—rare (see UC) *Bowel obstruction* *Fistula formation* *Perianal disease* *Malignancy*—large and small bowel cancer—5% 10y after diagnosis *Osteoporosis*
Psychological effects—chronic lifelong conditions which have major impact on work and domestic life. Self-help groups can be useful.	

Further information

NICE (2019) Crohn's disease: management. 🖱 www.nice.org.uk/guidance/ng129

NICE (2019) Ulcerative colitis: management. 🖱 www.nice.org.uk/guidance/ng130

Advice and support for patients

Crohn's and Colitis UK ☎ 0300 222 5700 🖱 www.crohnsandcolitis.org.uk

Irritable bowel syndrome

Irritable bowel syndrome (IBS) is a chronic (>6mo) relapsing and remitting condition of unknown cause with symptoms including: abdominal pain or discomfort; bloating; and change in bowel habit.

Diagnosis is clinical and there is no confirmatory test. Extremely common. Lifetime prevalence ≥20%, although ~50% never consult a GP. ♀ > ♂ (2.5:1). Symptoms can appear at any age.

Diagnosis of IBS Abdominal pain or discomfort that is:

- Relieved by defecation, or
- Associated with altered bowel frequency or stool form

And ≥2 of the following:

- Altered stool passage (straining, urgency, incomplete evacuation)
- Abdominal bloating (♀ > ♂), distension, tension or hardness
- Symptoms made worse by eating
- Passage of mucus

Other commonly associated symptoms Include lethargy, nausea, backache and bladder symptoms.

Differential diagnosis

- Colonic carcinoma
- Coeliac disease
- Inflammatory bowel disease (Crohn's disease or UC)
- Pelvic inflammatory disease
- Endometriosis
- GI infection
- Thyrotoxicosis

Investigation A diagnosis of exclusion. How far to investigate is a clinical judgement weighing risks of investigation against possibility of serious disease. Judgement is based on age of the patient, family history, length of history, and symptom cluster.

- **Patients <40y** Check FBC, CRP, ESR, and antibody testing to exclude coeliac disease (TTG/EMA)
- **Patients >40y** Colonic cancer must be excluded for any patient with a persistent, unexplained change in bowel habit—particularly towards looser stools (➔ p. 370)
- **Other investigations to consider**
    - Faecal calprotectin stool test ± referral for colonoscopy to exclude inflammatory bowel disease (as per local guidelines)
    - Thyroid function tests if other symptoms/signs of thyroid disease
    - Stool samples to exclude GI infection if diarrhoea
    - Endocervical swabs for *Chlamydia*
    - Laparoscopy to exclude endometriosis

Referral To gastroenterology/general surgery if (*U* = urgent; *S* = soon; *R* = routine):

- Passing blood (except if from an anal fissure or haemorrhoids)—*U*
- Abdominal, rectal or pelvic mass—*U*
- Unintentional/unexplained weight loss—*U/S*
- +ve faecal calprotectin, inflammatory markers, and/or anaemia—*U/S*
- >40y with new symptoms—*U* (if age >60y)*/S/R*
- Change in symptoms—especially if >40y—*U* (if age >60y)*/S/R*
- Atypical features (i.e. not those listed above)—*U/S/R*

- Family history of bowel or ovarian cancer—*R*
- Patient is unhappy to accept a diagnosis of IBS despite explanation—*R*

Treatment Reassure. Information leaflets are helpful. Encourage lifestyle measures, stress ↓, leisure time and regular physical exercise.

Diet Encourage patients to have regular meals and take time to eat. Avoid missing meals or leaving long gaps between eating.

- Drink ≤8 cups of fluid/d, especially water. Restrict tea/coffee to 3 cups/d. ↓ intake of alcohol and fizzy drinks
- ↓ intake of high-fibre foods (e.g. wholemeal/high-fibre flour and breads, cereals high in bran, and whole grains such as brown rice)
- ↓ intake of 'resistant starch' found in processed or re-cooked foods
- Limit fresh fruit to 3 × 80g portions/d
- For diarrhoea, avoid sorbitol, an artificial sweetener
- For wind and bloating consider ↑ intake of oats (e.g. oat-based breakfast cereal or porridge) and linseeds (≤1 tablespoon/d)
- Up to 50% may be helped by exclusion of certain foods. Diaries may help identify foods that provoke symptoms, e.g. dairy products, citrus fruits, caffeine, alcohol, tomatoes, gluten, and eggs. Refer to dietician

Specific measures

- Fibre/bulking agents Constipation-predominant IBS. Bran can make some patients worse. Oats and ispaghula husk are better tolerated. Laxatives are an alternative but avoid use of lactulose
- Antispasmodics e.g. mebeverine, peppermint oil. Limited effectiveness. If no response in a few days, switch to another—different agents suit different individuals. Once symptoms are controlled use prn
- Antidiarrhoeal preparations e.g. loperamide. Avoid codeine phosphate as may cause dependence. Use prn for patients with diarrhoea-predominant disease. Use pre-emptive doses to cover difficult situations (e.g. air travel)
- Antidepressants There is some evidence that low-dose amitriptyline, e.g. 10mg nocte or SSRIs may help symptoms
- Probiotics Some evidence of effectiveness. Consider a 4wk trial
- Psychotherapy and hypnosis Evidence of effectiveness but limited availability. Consider for cases that have failed to respond to first-line treatment after >1y
- FODMAPs diet Some evidence of effectiveness. Refer to dietician

Failure to respond to treatment Consider another diagnosis—review history and examination ± refer for further investigation.

Prognosis >50% still have symptoms after 5y.

Further information

NICE (2013) Faecal calprotectin diagnostic tests for inflammatory bowel disease. 🖱 www.nice.org.uk/guidance/dg11

NICE (2008, updated 2017) Irritable bowel syndrome in adults: diagnosis and management of irritable bowel syndrome in primary care. 🖱 www.nice.org.uk/guidance/cg61

Advice and support for patients

The IBS Network 🖱 www.theibsnetwork.org

Jaundice and abnormal liver function

Jaundice Yellow pigmentation of the tissues due to excessive bile pigment. Clinical jaundice appears when serum bilirubin >35 micromol/L.

Causes

- ↑ production of bilirubin (pre-hepatic) Haemolytic anaemia, drug-induced haemolysis, malaria, Gilbert's/Crigler-Najjar syndrome
- Defective processing (hepatic) Hepatitis, cirrhosis
- Blocked excretion (obstructive) Gallstones, pancreatic cancer, primary biliary cholangitis, primary sclerosing cholangitis, cholangiocarcinoma, sepsis, enlarged porta hepatis (e.g. 2° to lymphoma)

History Patients presenting with jaundice may have no other symptoms. General symptoms include tiredness, nausea, and pruritus. Ask about colour of the stools and urine (dark urine suggests conjugated hyperbilirubinaemia and hepatobiliary disease). Check alcohol consumption.

Examination Mild jaundice is best seen by examining the sclerae in natural light. Look for signs of chronic liver disease and examine the abdomen for masses and hepatomegaly.

Investigations Initially check FBC and liver function tests—Table 12.17. Further investigations depend on the results.

Management Treat the cause of the jaundice. Most patients (except those with Gilbert's syndrome or self-limiting viral hepatitis) will require specialist referral. Refer patients with pre-hepatic/hepatic jaundice to a hepatologist; refer patients with post-hepatic jaundice to a hepatobiliary surgeon. If aged ≥40y + jaundice—refer urgently to be seen in <2wk.

Neonatal jaundice ➔ p. 846

Abnormal liver function

Raised AST/ALT/GGT in isolation Liver enzymes can ↑ transiently as a result of viral infection, drugs, or alcohol. ALT tends to be more raised in viral and autoimmune hepatitis; AST tends to be more raised in patients with fatty liver; raised GGT is associated with alcohol excess.

- Check medication including herbal medicines
- Stop alcohol
- Repeat LFTs:
  - In 1mo if AST/ALT are <3× upper limit of normal
  - In 1wk if AST/ALT are ≥3× upper limit of normal
- If still raised request hepatitis screen and USS, or refer to hepatology or gastroenterology depending on clinical state
- If ALT is <2× upper limit of normal and hepatitis screen is negative:
  - If USS is normal, repeat LFTs every 3–6mo to see if abnormalities settle—may be due to drugs, alcohol, or early fatty liver disease
  - If USS shows fatty liver and consuming excess alcohol—advise abstinence from alcohol and recheck LFTs every 3–6mo
  - If USS shows fatty liver and alcohol consumption is within normal limits, advise weight ↓ and low-fat diet, treat metabolic syndrome, and recheck LFTs every 3–6mo
- In all other cases, refer for specialist opinion

Table 12.17 Distinguishing different types of jaundice

	Type of jaundice		
	Pre-hepatic	Hepatic	Cholestatic
Tests			
Bilirubin	↑↑	↑↑	↑↑
AST/ALT	Normal	↑↑	↑
Alkaline phosphatase	Normal	↑	↑↑↑
Hb	↓	Normal	Normal
Jaundice	Mild, lemon yellow	May be marked jaundice	May be marked jaundice
Other symptoms	Urine is *not* darkened	Tender, enlarged liver	Enlarged liver, itching skin, pale stools

❗ A mixed picture is common and can be confusing.

Raised bilirubin Possible causes include:

- Hepatitis/biliary obstruction—if ALT/AST are ↑, refer for liver USS and do a hepatitis screen or refer as for jaundice
- Haemolysis—check FBC/reticulocyte count—refer to haematology if abnormal
- Gilbert's disease—likely if serum bilirubin is <40mmol/L and ALT/AST and RBC/reticulocytes are normal. Bilirubin levels ↑ after a fast
- If isolated ↑ bilirubin >40mmol/L—probably still Gilbert's syndrome but refer

Raised alkaline phosphatase Usually originates from liver or bone. Bone is more likely if serum Ca^{2+} and phosphate are raised and GGT is normal. ↑ may be associated with any liver disease but is particularly marked in patients with biliary obstruction and primary biliary cholangitis.

Medications that cause raised AST/ALT

- Most penicillins (especially co-amoxiclav) and minocycline
- Antifungals
- Statins
- Antiepileptics
- NSAIDs
- Some herbal medicines
- Some recreational drugs

Statins and abnormal liver function Statins cause a biochemical abnormality, but do not cause liver failure and are not contraindicated in compensated liver disease. May improve fatty liver disease. Measure liver function tests pre-treatment and after 1–3mo. Thereafter measure each 6mo for 1y. Discontinue if AST/ALT ↑ and stays at >3× normal.

Hepatitis screen Check as appropriate:

- Hepatitis A, B, and C serology
- EBV serology
- Liver autoantibodies
- Iron studies and transferrin saturation to exclude haemochromatosis
- α1 antitrypsin level
- Serum copper and caeruloplasmin levels (if <40y)
- AFP
- Fasting blood glucose and lipids
- HbA1c

Fatty liver disease

Fatty liver describes the pathological process where fat is deposited in liver cells, but there is no inflammation or scarring. Common condition affecting up to 1:4 adults in the UK.

Presentation

- Usually asymptomatic. >50% present after investigation for abnormal LFTs (➲ p.390). Characteristic 'bright' appearance on liver USS
- Less frequently presents with a smoothly enlarged liver or symptoms—nausea, vomiting, abdominal pain, fat embolus (may be fatal)

Broadly divides into 2 forms:

Alcohol-associated fatty liver disease (AFLD) Defined as fatty liver disease associated with daily ethanol consumption >20g (♀) or 30g (♂). Earliest stage of alcohol-related liver disease. Affects ≈90% of people who drink more than recommended limits. Can develop in weeks.

Blood markers (may be normal)

- Typically serum AST and ALT are ↑, but serum AST >serum ALT
- GGT may be ↑
- Alkaline phosphatase may also be ↑ but usually <2× upper limit of normal

Primary care management

- Identification of hazardous/harmful drinking + brief interventions to ↓ consumption—➲ p. 158
- Referral to specialist alcohol addiction service if needed
- If persistent heavy drinker (♀ >35U/wk; ♂ >50U/wk) refer for transient elastography (TE) to assess for fibrosis/cirrhosis. Refer for specialist management if advanced fibrosis/cirrhosis. If TE is not available, follow local protocols for assessment of fibrosis, Alternatives include: serum fibrosis markers, FIB-4 test, fibrotest, AST:ALT ratio, and AST to Platelet Ratio Index (APRI). Repeat every 2y
- Consider thiamine supplements if chronic alcohol dependence: if severe, 200–300mg/d; if mild, 10–25mg/d

Prognosis AFLD is fully reversible and can resolve in <6wk with complete abstinence from alcohol. If excessive alcohol use continues, predisposes to alcoholic hepatitis and cirrhosis (irreversible).

Alcohol misuse ➲ p. 158

Chronic hepatitis ➲ p. 394–5

Cirrhosis ➲ p. 396

Non-alcoholic fatty liver disease (NAFLD)[N] Very common. Affects 20–30% of the UK population. Prevalence has ↑ ×2 over the past 20y. Associated with obesity, insulin resistance and metabolic syndrome (➲ p. 313).

Screening With liver USS is not recommended except for children/young people with metabolic syndrome/type 2 DM—repeat every 3y.

Blood markers (may be normal)

- Typically, serum AST and ALT are ↑ but serum ALT >serum AST
- GGT may be ↑
- Alkaline phosphatase may be ↑ but usually <2× upper limit of normal

Complications

- A high proportion develop DM long term
- More severe disease results in non-alcoholic steato-hepatitis (NASH)—12% with NASH develop cirrhosis

Assessment for fibrosis Once NAFLD has been diagnosed, assess for fibrosis risk using the Enhanced Liver Fibrosis (ELF) test, if available. Alternatives include: FIB-4 test, NAFLD score, or TE.

Referral for specialist assessment

- Child/young person with suspected NAFLD on USS
- Any age with significant liver fibrosis risk (e.g. ELF score >10.51)

Management in primary care

- Lifestyle advice—low-fat diet, weight ↓, ↑ exercise, ↓ alcohol
- Consider referral for bariatric surgery if meets local referral criteria
- Repeat ELF or other fibrosis score every 3y

Drug treatment

- Not usually indicated in primary care—in secondary care, treatment (unlicensed) with metformin, pioglitazone and/or vitamin E may be considered
- Treatment with statins is ineffective

Other rarer causes of fatty liver disease

- Drugs, e.g. amiodarone, tamoxifen, valproate
- Inflammatory bowel disease
- Malnutrition
- Pregnancy

Further information

NICE (2016) Cirrhosis in over 16s. 🖱 www.nice.org.uk/guidance/ng50

NICE (2016) Non-alcoholic fatty liver disease: assessment and management. 🖱 www.nice.org.uk/guidance/ng49

Hepatitis

Acute hepatitis May be asymptomatic or present with fatigue, flu-like symptoms, fever, light stools, dark urine, and/or jaundice. *Causes*:

- Viral hepatitis (e.g. HBV, HAV, EBV)
- Alcohol (➔ p. 158)
- Drugs (e.g. diclofenac, co-amoxiclav)
- Toxins
- Obstructive jaundice
- Other infections—malaria, Q fever, leptospirosis, yellow fever

Management Check LFTs, FBC, U&E, eGFR, hepatitis serology. Treat the cause. Admit if condition is poor or rapidly deteriorating; refer for investigation if sustained abnormalities in liver function with unclear cause (➔ p. 390). *Complications*: chronic hepatitis, acute liver failure.

Hepatitis A (HAV) Common. *Spread*: faecal–oral route. Patients are infectious 2wk before feeling ill. Incubation is 2–7wk (average 4wk). *High-risk groups*: travellers to high-risk areas, institutional inhabitants and workers, IV drug abusers, patients with high-risk sexual practices. May be asymptomatic (especially young children) or present with fever, malaise, fatigue, anorexia, nausea/vomiting, abdominal pain, diarrhoea, tender hepatomegaly, pale stools, dark urine, and/or jaundice (70–80% adults).

Management Check LFTs (hepatic jaundice—Table 12.17, ➔ p. 391) and hepatitis serology. IgM antibodies signify recent infection. Immunity follows infection, and IgG remains detectable lifelong. Management is supportive. Avoid alcohol until LFTs are normal. Most recover in <2mo. Hepatitis A does not cause chronic liver disease; there is no carrier state.

Prevention Vaccination is indicated for travellers to high-risk areas, people with chronic liver disease, or those working in high-risk situations. Preparations available include monovalent vaccine (e.g. Havrix®), hepatitis A and B combined vaccine (Twinrix®), and hepatitis A and typhoid combined vaccine (e.g. Hepatyrix®). Passive immunization with human immunoglobulin gives protection for ≤3mo and is used for short-term travel or protecting household contacts of sufferers.

Hepatitis E (HEV) Similar to HAV infection. Usually acquired in developing countries. Incubation is 2–9wk (average 40d). Diagnosis is made with serology. Treatment is supportive. There is no chronic state. Mortality in pregnancy can be as high as 20%. No vaccine exists.

Hepatitis B (HBV) ➔ p. 718

Hepatitis C (HCV) ➔ p. 718

Chronic hepatitis Hepatitis lasting >6mo. May be asymptomatic or present with fatigue; RUQ pain; jaundice; arthralgia; signs of chronic liver disease—gynaecomastia, testicular atrophy, clubbing, palmar erythema, leuconychia, peripheral oedema, spider naevi, portal hypertension, recurrent infection; and/or complications—acute liver failure, cirrhosis, hepatocellular carcinoma. *Causes*:

- Viral hepatitis
- Alcohol—➔ p. 158
- Drugs (e.g. nitrofurantoin, methyldopa, isoniazid)
- Chronic autoimmune hepatitis
- Primary biliary cholangitis
- Wilson's disease
- Haemochromatosis
- α1-antitrypsin deficiency
- Sarcoidosis

Management Check LFTs; FBC; U&E; eGFR; and hepatitis screen (➔ p. 391). Refer for specialist care.

Chronic autoimmune hepatitis Also known as '*chronic active hepatitis*'. Typically young women. Associated with personal/family history of autoimmune disease (e.g. RA, vitiligo). Diagnosis is confirmed with liver biopsy and autoimmune markers. Specialist management is with steroids ± immunosuppressants.

Primary biliary cholangitis Slow progressive cholangio-hepatitis eventually resulting in cirrhosis. ♀:♂ ≈9:1. *Peak age at presentation*: 45y. *Cause*: probably autoimmune. *Associations*:

- Thyroid disease
- Sjögren's syndrome
- CREST syndrome
- Coeliac disease
- Hepatic and extra-hepatic malignancy
- Pancreatic hyposecretion

Presentation 50% are asymptomatic at presentation. *Symptoms/signs*:

- Fatigue
- Pruritus
- Arthralgia
- Osteoporosis/osteomalacia
- Hirsutism
- Obstructive jaundice (late)
- Symptoms/signs of cirrhosis or liver failure

Investigation and management *Blood*: LFTs (↑ alk phos, ↑ALT, ↑GGT). Liver biopsy is diagnostic. Refer for specialist care. If asymptomatic, 1:3 remain symptom free—the rest develop symptoms in 2–4y. Median survival is 7–10y. Liver transplant is an option. Prognosis following transplant is good but recurrence may occur in the transplanted liver.

Information and support for patients

British Liver Trust ☎ 0800 652 7330 🖱 www.britishlivertrust.org.uk
PBCers Organization 🖱 www.pbcers.org
Wilson's Disease Association 🖱 www.wilsonsdisease.org

Liver failure and portal hypertension

⚠ **Acute liver failure** Presents with sudden onset of severe illness.

- Jaundice
- Hypoglycaemia
- Hepatic encephalopathy—ranges from mild confusion and irritability through drowsiness and ↑ confusion to coma
- Haemorrhage—due to deranged clotting factors
- Ascites—hepatosplenomegaly and ascites are not usually prominent
- Infection
- Nausea ± vomiting
- ↑ BP
- Fetor hepaticus (sweet smell on the breath)

Causes

In previously healthy patients:

- Viral hepatitis
- Weil's disease
- Paracetamol overdose
- Halothane
- Idiosyncratic drug reactions
- Fungal/plant toxins
- Malignant infiltration
- Chemical exposure (e.g. carbon tetrachloride)
- Heatstroke
- Budd–Chiari syndrome
- Pregnancy
- Wilson's disease
- Reye's syndrome

In patients with chronic liver disease:

- Infection
- GI bleeding
- Sedation
- Diuretics and/or electrolyte imbalance
- Alcohol binges
- Constipation

Management Admit as emergency unless an expected terminal event. Prognosis is poor (<60% survive).

Cirrhosis The liver is replaced by fibrotic tissue and regenerating nodules of hepatocytes.

Common causes/risk factors

- Hepatitis B infection ➔ p. 718
- Alcohol misuse ➔ p. 158
- Hepatitis C infection ➔ p. 718
- Type 2 DM or BMI ≥30kg/m^2

Screening high-risk groups Offer transient elastography (TE) if:

- Chronic hepatitis B/C infection—retest every 2y or more frequently according to specialist advice
- Persistent heavy drinker (♀ >35U/wk; ♂ >50U/wk) or confirmed alcohol-related liver disease—retest every 2y
- Non-alcoholic fatty liver disease + advanced liver fibrosis (Enhanced Liver Fibrosis test score ≥10.51)—retest every 3y
- Liver biopsy is an alternative if TE is not suitable

❗ Do not use liver function blood tests to rule out cirrhosis.

Rarer causes

- Chronic active hepatitis ➔ pp. 394–5
- Primary biliary cholangitis ➔ p. 395
- α1-antitrypsin deficiency ➔ p. 398
- Wilson's disease ➔ p. 398
- Budd–Chiari syndrome ➔ p. 399
- Haemochromatosis ➔ p. 398

Presentation Variable. May be an incidental finding with no symptoms or may present with non-specific symptoms, e.g. weakness, fatigue, lethargy. Specific symptoms/signs include:

- Hepatomegaly (but liver becomes small and hard in late stages)
- Spider naevi
- Dupuytren's contracture
- Palmar erythema
- Gynaecomastia
- Testicular atrophy
- Clubbing
- Xanthelasma/xanthomata
- Portal hypertension
- Splenomegaly

Late signs Occur when the liver can no longer compensate for the damage to it—jaundice, hepatic encephalopathy, leuconychia, and oedema (due to hypoalbuminaemia).

Management

- Refer to gastroenterology/hepatology for expert advice
- Treat the cause where possible
- Avoid alcohol completely and refer to dietician for advice on nutrition
- Pruritus 2° to jaundice may respond to cholestyramine
- Give pneumococcal vaccination and annual influenza vaccination

Complications

- Portal hypertension (± bleeding oesophageal varices)
- Encephalopathy
- Hepatocellular carcinoma
- Ascites (bacterial peritonitis complicates 1 in 4 cases—consider prophylaxis with ciprofloxacin)
- Renal failure

Prognosis Very variable—depending on age, cause, and willingness to modify contributing lifestyle factors.

Portal hypertension Portal venous pressure is raised due to obstruction of the portal system before, within, or after the liver. In Western countries the most common cause is cirrhosis.

- Elevated portal venous pressure → collaterals between the portal and systemic circulation (including oesophageal varices). Usually presents with haematemesis and/or melaena from bleeding varices
- Ascites develops if there is coexistent liver failure with hypoproteinaemia and hyperaldosteronism
- Splenomegaly is common → thrombocytopenia and leucopenia
- *Signs*: splenomegaly (80–90%), ascites, dilated veins around the umbilicus (rare), purpura, signs of chronic liver disease
- After a diagnosis of cirrhosis, offer upper GI endoscopy to detect oesophageal varices—repeat every 3y if –ve
- Refer to gastroenterology/hepatology. Specialist management is essential. If GI bleeding, refer as a 'blue light' emergency

Hepatocellular carcinoma ➔ p. 398

Further information

NICE (2016) Cirrhosis in over 16s. 🖱 www.nice.org.uk/guidance/ng50

Information and support for patients

British Liver Trust ☎ 0800 652 7330 🖱 www.britishlivertrust.org.uk

Other liver disease

Gilbert's syndrome Inherited metabolic disorder causing unconjugated hyperbilirubinaemia. *Prevalence*: ~1–2%. Onset is shortly after birth—but the condition may go unnoticed for years. Jaundice occurs during intercurrent illness. ↑ bilirubin on fasting can confirm the diagnosis. Liver biopsy is normal. No treatment is required and prognosis is excellent.

Primary haemochromatosis Autosomal recessive condition of excess gut absorption of iron → iron deposition and damage to heart, liver, pancreas, joints, and pituitary. ~1:400 people are homozygous for the condition but expression is highly variable. ♂ > ♀ (♀ present ~10y later). Often an incidental finding, or found by screening relatives of affected individuals (genetic testing or serum ferritin). *Symptoms/signs*:

- Tiredness
- Arthralgia/arthritis
- Skin pigmentation
- Hepatomegaly ± signs of cirrhosis
- DM
- Impotence/testicular atrophy
- Cardiomyopathy

Investigation and management Blood: ↑ ferritin; ↑ iron; transferrin saturation >70%; total iron binding capacity ↓. Refer. Liver biopsy is diagnostic. Venesection returns life expectancy to normal.

Secondary haemochromatosis Iron overload from frequent transfusions, e.g. for haemolysis. Specialist management with chelation therapy to ↑ iron excretion is required.

α1-antitrypsin deficiency Autosomal recessive disorder. Defective α1-antitrypsin production → lung, and more rarely liver damage. 💣 Treatment with IV α1-antitrypsin ↓ progression of COPD. Paracetamol may protect the liver. Encourage use for minor illness. Liver transplantation may eventually be needed.

Wilson's disease (hepatolenticular degeneration) Rare, autosomal recessive disorder. Defective biliary copper excretion → accumulation of copper in the liver, brain, kidney, and cornea. Treatment is with penicillamine. Liver transplantation is the only treatment if presentation is with acute liver failure.

Benign tumours Hepatomegaly ± RUQ pain or an incidental finding.

Common types Hepatic adenoma, fibroma, leiomyoma, lipoma, haemangioma, focal nodular hyperplasia (e.g. with cirrhosis).

Management Refer to gastroenterology to exclude malignancy and confirm diagnosis. Urgency depends on clinical picture and USS findings.

Hepatocellular cancer (HCC) Rare in the UK (100 new cases and 100 deaths/y). Much more common in areas of the world where hepatitis B is endemic (e.g. China, India). Usually arises from regenerating nodules in a cirrhotic liver. *Peak age*: 60–70y. Intra- and extrahepatic spread is common and occurs early.

Surveillance All patients with confirmed cirrhosis should have regular USS surveillance[N]. Usually this is specialist led.

Presentation In a patient with known cirrhosis:

- Fatigue
- Fever
- Anorexia and/or weight ↓
- Ascites
- Rapid deterioration in liver function
- Haemorrhage into the peritoneal cavity (often fatal)
- *Budd–Chiari syndrome* (occlusion of the hepatic vein resulting in jaundice, epigastric pain, and shock)
- *Examination*—may reveal an abdominal mass, hepatomegaly ± an arterial bruit over the tumour

Management If suspected, check AFP and refer for urgent assessment. AFP >500ng/mL in a patient with known cirrhosis is almost certainly diagnostic. The most important prognostic factors are the number and size of the liver lesions and the presence of vascular involvement. 95% of patients with cirrhosis have disease too extensive for curative surgery, or their severely compromised liver function makes radical surgery inappropriate. 50% of patients without cirrhosis have resectable tumours. Surgery may be combined with liver transplantation. Inoperable tumours may be treated with hepatic artery ligation or embolization. Tumours respond poorly to chemo- or radiotherapy.

Overall prognosis Patients with cirrhosis—median survival 3mo; patients without cirrhosis—median survival 1y.

Cholangiocarcinoma Rare adenocarcinoma of the biliary tract. May be associated with UC. Typically presents in patients >60y with jaundice, RUQ pain and weight loss. The only effective treatment is surgery, which is only possible in ~10–20% of patients. Selected fit patients with unresectable disease may be offered palliative chemotherapy or enrolment in a clinical trial. Median survival 4–6mo.

Secondary tumours The most common type of liver tumours—usually signalling late disease. *Presentation*: hard, enlarged, knobbly liver ± RUQ pain ± jaundice (late). If found and no history of malignancy refer to oncology/general surgery for urgent referral to find the primary.

Primary tumours commonly metastasizing to the liver Lung, breast, large bowel, stomach, uterus, pancreas, carcinoid, lymphoma, leukaemia.

Further information

British Society for Haematology 🖱 www.b-s-h.org.uk/guidelines/

- Investigation and management of a raised serum ferritin (2018).
- Diagnosis and therapy of genetic haemochromatosis (2018).

NICE (2015, updated 2017) Suspected cancer: recognition and referral 🖱 www.nice.org.uk/guidance/ng12

Advice and support for patients

Alpha-1 Support for people with α1-antitrypsin deficiency. 🖱 www.alpha1.org.uk
British Liver Trust ☎ 0800 652 7330 🖱 www.britishlivertrust.org.uk
Cancer Research UK ☎ 0808 800 4040 🖱 www.cancerhelp.org.uk
Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk
Wilson's Disease Association 🖱 www.wilsonsdisease.org

Gallbladder disease

Gallstones Gallstones are increasingly common. 9% of 60y-olds have them and prevalence ↑ with age.

Other risk factors

- Gender (♀ > ♂)
- Body weight—prevalence ↑ with weight; also associated with rapid weight ↓
- Race—in the USA Native American > Hispanic > white > black
- Affluency
- Pregnancy (and possibly HRT but not COC pill)
- Alcohol is protective
- Diet—vegetarian diet is protective

Associated conditions

- Haemolysis
- DM
- Hypertriglyceridaemia
- Cirrhosis
- Crohn's disease
- Partial gastrectomy

Drugs which cause gallstones Clofibrate (and other fibric acid derivatives); octreotide (somatostatin analogue).

Presentation Gallstones are blamed for many digestive symptoms—they are probably innocent in most cases. 70% of stones in the gallbladder do not cause symptoms. Common presentations—Table 12.18.

Management of gallstones

- Advise the patient to stick to a low-fat diet
- Refer for surgical review ± further evaluation (e.g. ERCP—endoscopic retrograde cholangiopancreatography)
- Gallstones can be removed by cholecystectomy (laparoscopic or open) or ERCP or may be dissolved with ursodeoxycholic acid (stones <5mm diameter—40% recur in <5y) or shattered with lithotripsy (1 in 3 develop biliary colic afterwards)
- Persistent digestive symptoms after surgery are common (50% after cholecystectomy) and difficult to treat

Gallbladder cancer Rare. ♀ > ♂. Gallstones are a predisposing factor. Typically presents in patients >40y with RUQ pain, anorexia, weight ↓, and jaundice. Refer for specialist upper GI assessment (to be seen in <2wk) if suspected. Surgical resection offers the only hope of cure but disease is usually advanced at presentation. Selected fit patients with unresectable disease may be offered palliative chemotherapy or enrolment in a clinical trial. Prognosis is poor.

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Information and support for patients

British Liver Trust ☎ 0800 652 7330 🖱 www.britishlivertrust.org.uk

Table 12.18 Presentation and management of gallstone disease

	Presentation	Management
Biliary colic	Clear-cut attacks of severe upper abdominal pain which may radiate → back/shoulder tip, lasting ≥½ h and causing restlessness ± jaundice ± nausea or vomiting. *Examination:* tenderness ± guarding in the right upper quadrant (↑ on deep inspiration—Murphy's sign)	*Treat* acute attacks with pethidine (50mg IM/po) or diclofenac (50–100mg IM/PO/PR) + prochlorperazine 12.5mg IM or domperidone 10mg po/PR for nausea *Admit if*: uncertain of diagnosis, inadequate social support, persistent symptoms despite analgesia, suspicion of complications, and/or concomitant medical problems (e.g. dehydration, pregnant, DM, Addison's) *Investigate* for gallstones with abdominal USS to prove diagnosis when settled *Differential diagnosis*: any cause of acute abdomen *Treat* gallstones to prevent recurrence
Acute cholecystitis/ cholangitis	Pain and tenderness in the right upper quadrant/ epigastrium ± vomiting *Examination*: tenderness ± guarding in the right upper quadrant ± fever ± jaundice	*Treatment*: broad-spectrum antibiotic (e.g. ciprofloxacin) and analgesia as for biliary colic *Admit if*: generalized peritonism, diagnosis uncertain, very toxic, concomitant medical problems (e.g. dehydration, DM, Addison's, pregnancy), inadequate social support, or not responding to medication *Empyema* occurs when the obstructed gallbladder fills with pus. Presents with persistent swinging fever and pain. Usually requires cholecystectomy ± surgical drainage *Investigate and follow-up* to prevent recurrence as for biliary colic
Pancreatitis	➔ p. 402	➔ p. 402
Gallstone ileus	Occurs usually after an attack of cholecystitis. A stone perforates from the gallbladder into the duodenum and impacts in the terminal ileum causing bowel obstruction	➔ p. 372
Chronic cholecystitis	Vague intermittent abdominal discomfort, nausea, flatulence and intolerance of fats	*Investigate* for gallstones with abdominal USS to prove the diagnosis *Differential diagnosis*: reflux, IBS, upper GI tumour, PU Refer for treatment of gallstones
Jaundice	Cholestatic jaundice—➔ p. 390 ± right upper quadrant pain	➔ p. 390

Pancreatitis

Acute pancreatitis Premature activation of pancreatic enzymes results in autodigestion and tissue damage. Most episodes are mild and self-limiting but 1:5 patients have a severe attack. Overall mortality ≈5–10%. May be recurrent.

Causes In 10% patients no cause is identified.

- Common causes (80%) Gallstones, alcohol
- Rarer causes
  - Drugs (e.g. azathioprine)
  - Trauma
  - Pancreatic tumours
  - Post-ERCP
  - Viral infection (mumps, HIV, Coxsackie B)
  - Mycoplasma infection
  - Hypercalcaemia
  - Hyperlipidaemia
  - Pancreas divisum (normal variant in 7–8% of the white population)
  - Familial pancreatitis
  - Vasculitis
  - Ischaemia or embolism
  - Pregnancy
  - End-stage renal failure

Presentation

- Poorly localized, continuous, boring epigastric pain which ↑ over ~1h—often worse lying down ± radiation to the back (50%)
- Nausea ± vomiting

Examination

- General Tachycardia, fever, shock, jaundice
- Abdominal Localized epigastric tenderness or generalized abdominal tenderness; abdominal distension ± ↓ bowel sounds; evidence of retroperitoneal haemorrhage (periumbilical and flank bruising—rare)

Management Admit as an acute surgical emergency. Prior to transfer give analgesia with pethidine (morphine may induce spasm of the sphincter of Oddi).

Complications Delayed complications may present in general practice—suspect if persistent pain or failure to regain weight or appetite. Complications include:

- Pancreatic necrosis
- Pseudocyst—localized collection of pancreatic secretions
- Fistula/abscess formation
- Bleeding or thrombosis

Prevention of further attacks

- Avoid factors that may have caused pancreatitis, e.g. alcohol, drugs
- Advise patients to follow a low-fat diet
- Treat reversible causes e.g. hyperlipidaemia, gallstones

Chronic pancreatitis Chronic inflammation of the pancreas results in gradual destruction and fibrosis of the gland ± loss of pancreatic function → malabsorption and DM.

Cause Alcohol is responsible for most cases. *More rarely*: familial; CF; haemochromatosis; pancreatic duct obstruction (gallstones/pancreatic cancer); hyperparathyroidism.

Presentation

- Constant or episodic epigastric pain radiating to the back and relieved by sitting forwards
- Vomiting
- Weakness
- Jaundice
- Steatorrhoea
- Weight ↓
- DM
- Chronic poor health

Management Refer to gastroenterology. *Treatment*:

- **Diet** Low-fat, high-protein, high-calorie diet with fat-soluble vitamin supplements. Refer to dietician
- **Pancreatic enzyme supplementation** e.g. Creon® capsules pre-meals. May improve diarrhoea
- **Alcohol abstinence**
- **Pain control** Provide analgesia—beware of opioid abuse. Consider referral for coeliac plexus block
- **Surgery** Pancreatectomy or pancreaticojejunostomy for pancreatic duct stricture, obstructive jaundice, unremitting pain, or weight loss
- **Diabetes management**

Pancreatic insufficiency Global ↓ function of the pancreas. *Causes*:

- **Child** Cystic fibrosis
- **Adult** Chronic pancreatitis, pancreatic tumour, pancreatectomy, total gastrectomy

Presentation Malabsorption (frequent loose, odorous stools ± abdominal pain), weight loss or failure to thrive, DM.

Management Take specialist advice. Treat the underlying cause. Treat associated DM. Supplement digestive enzymes (e.g. with Creon).

Pancreatic tumours

Pancreatic cancer accounts for 3% of all malignancies causing ~7800 deaths/y in the UK. 80% of cases occur in patients >60y. ♂ > ♀ (3:2).

Risk factors

- Smoking—causes 25–30% of pancreatic cancers in the UK. Risk returns to non-smoker levels 10–20y after cessation
- Chronic pancreatitis—usually related to excess alcohol
- Type 2 (non-insulin dependent) DM—relative risk ≈1.8
- Obesity—↑ risk by 19%
- Genetic—5% pancreatic cancers are hereditary—characterized presentation aged <30y and +ve FH
- Occupation—cancer is ↑ among nickel workers, and workers exposed to insecticides, radiation, lead, iron, or chromium

Tumour characteristics

- The majority of pancreatic tumours develop in the exocrine part of the gland. 95% of tumours are adenocarcinomas. Rarely tumours develop from the endocrine part—these have better prognosis
- 75% arise in the head of the pancreas, 15% from the body, and 10% from the tail. Tumours arising in the head of the pancreas tend to present earlier and are easier to remove
- Spread to local LNs occurs early and metastatic spread to the peritoneum, liver, and lungs is frequently found at presentation

Presentation Non-specific with:

- Gradual deterioration in health or fatigue
- Anorexia or weight ↓;
- Pain—epigastric ± radiation → back—may be relieved by sitting forward
- Diarrhoea/steatorrhoea due to malabsorption
- Early satiety, dyspepsia, or nausea/vomiting (gastric outlet obstruction)
- Obstructive jaundice
- Pancreatitis
- New DM
- Spontaneous venous thrombosis

Examination Check for weight ↓, epigastric or left upper quadrant mass, hepatomegaly, jaundice. If jaundice is present the gallbladder may be palpable as a small rounded mass beneath the liver.

Primary care management Refer for urgent assessment (in <2wk)^{N}.

To a team specializing in upper GI cancer if:

- Upper abdominal mass consistent with pancreatic cancer
- Aged ≥40y + jaundice

For direct access CT (or USS if CT not available) if ≥60y + weight loss AND ≥1 of:

- Diarrhoea
- Back pain
- Abdominal pain
- Nausea/vomiting
- Constipation
- New-onset diabetes

For direct access USS if Upper abdominal mass consistent with enlarged liver or gallbladder.

Specialist management Diagnosis is confirmed using a combination of USS, CT, MRI, and/or ERCP. The only potentially curative treatment is surgery but <15% of patients are suitable for surgery at presentation. The operation of choice is a Whipple's procedure (pancreaticoduodenectomy). Surgery is associated with significant morbidity; mortality 5–15%.

Prognosis Those undergoing surgical resection have 5y survival of 7–25% (median survival 11–20mo) but those that survive 5y are likely to survive long-term. Median survival for those with irresectable locally advanced disease is 6–11mo, and 2–6mo if metastatic disease.

Palliative treatment Patients with locally advanced/metastatic disease may benefit from surgical bypass of common bile duct and/or duodenal obstruction. An alternative is a biliary stent. Chemotherapy may give some survival benefit. Refer for palliative care support early.

Endocrine tumours In all cases specialist management is required:

Glucagonoma Islet cell tumour of the pancreas. Most are malignant and 90% have liver or LN metastases at presentation. 5–20% of tumours occur as part of multiple endocrine neoplasia (MEN-I) syndrome. *Presents with:*

- Attacks of hyperglycaemia (DM in >50%)
- Skin changes—sore mouth, necrolytic migratory erythema (70%—rash which starts as an erythematous rash then blisters before crusting)
- Weight ↓/cachexia (60%)
- Tendency to venous thrombosis (11%)
- Anaemia
- Diarrhoea
- Depression/psychosis

Insulinoma Tumour of the APUD cells of the Islets of Langerhans. >90% are benign. 7–8% are associated with MEN-I syndrome. Presents with episodes of hypoglycaemia, especially when exercising or fasting. ↑ appetite and frequent food intake to avoid hypoglycaemia often results in substantial weight gain.

Somatostatinoma Uncommon islet cell tumour. Most are large tumours (>5cm) in the head/body of the pancreas. Presents with gallstones, steatorrhoea, and DM.

❗ Extrapancreatic somatostatinomas can present in association with neurofibromatosis type I and phaeochromocytoma.

Verner Morrison syndrome An intestinal vasointestinal peptide (VIP) producing tumour results in profuse watery diarrhoea → dehydration, metabolic acidosis, and ↓ K^+. Also associated with insulin resistance and impaired glucose tolerance. VIPomas account for <10% islet cell tumours. 60% are malignant.

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral.
🖱 www.nice.org.uk/guidance/ng12

Advice and support for patients

Cancer Research UK ☎ 0808 800 4040 🖱 www.cancerhelp.org.uk
Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk

Chapter 13

Renal medicine and urology

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Laboratory tests

Serum creatinine (Cr) Commonly ordered test to detect renal dysfunction. Can be used to calculate estimated glomerular filtration rate (eGFR) when corrected for age, sex, weight, and ethnicity (➔ p. 410). ↓ in GFR is associated with ↑ in serum Cr.

Urea Commonly ordered test to detect renal dysfunction. While a ↓ in GFR is associated with an ↑ in serum urea, serum urea may also vary independently of the GFR. Causes of abnormal serum urea:

- **↑ serum urea** (>6.7mmol/L) Renal failure, GI bleeding, high-protein diet, drugs (e.g. high-dose steroids, tetracycline), dehydration
- **↓ serum urea** (<2.5mmol/L) Liver disease (↓ urea production), anabolic state, high ADH levels (high GFR), starvation or low-protein diet, pregnancy

⚠ Plasma potassium >6.5mmol/L needs urgent treatment.

- Check it is not an artefact, e.g. due to haemolysis inside the bottle
- Admit for investigation of cause and treatment

Hyperkalaemia High serum potassium (>5mmol/L). *Causes*: Table 13.1. Treat the cause.

ECG changes associated with hyperkalaemia Tall, tented T waves; small P wave; wide QRS complex becoming sinusoidal, VF.

Hypokalaemia Low serum potassium (<3.5mmol/L). Presents with muscle weakness, hypotonia, cardiac arrhythmias, cramps, and tetany. *Causes*: Table 13.1. If K^+ >2.5mmol/L and no symptoms, give oral potassium supplement. ❗ If the patient is taking a thiazide diuretic, hypokalaemia >3.0mmol/L rarely needs treating.

⚠ Plasma potassium <2.5mmol/L needs urgent treatment—admit.

ECG changes associated with hypokalaemia Small/inverted T waves; prominent U wave; prolonged P–R interval; depressed ST segment.

Hyponatraemia Low serum sodium (<135mmol/L). Rarely symptomatic in general practice. May present with signs of water excess—confusion, fits, ↑ BP, cardiac failure, oedema, anorexia, nausea, muscle weakness. *Causes*: Table 13.1. *Management*: treat the cause. If unwell admit for investigation.

Hypernatraemia Excess serum sodium (>145mmol/L). Rare in general practice. *Presentation*: thirst, confusion, coma, fits, signs of dehydration—dry skin, ↓ skin turgor, postural hypotension, and oliguria if water deficient. *Causes*: Table 13.1. *Management*: admit for investigation.

Glycosuria ➔ p. 312

Proteinuria Excess protein in the urine. Risk factor for renal disease and CVD. Usually renal in origin. Urine dipsticks are not sensitive for low levels of proteinuria/albuminuria. If proteinuria on dipstick, exclude UTI and repeat. If persists, check dipstick for haematuria, albumin:creatinine ratio (ACR) or protein:creatinine ratio (PCR), serum Cr, and BP.

Table 13.1 Causes of altered serum electrolytes

↑ potassium (>5mmol/L)	↓ potassium (<3.5mmol/L)	↑ sodium (>145mmol/L)	↓ sodium (<135mmol/L)
Renal failure Drugs, e.g. ACE inhibitors, excess K^+ therapy, K^+-sparing diuretics Addison's disease Metabolic acidosis (DM) Artefact (haemolysed sample)	Diuretics Cushing's syndrome/steroids Vomiting and/or diarrhoea Conn's syndrome Villous adenoma of the rectum Purgative or liquorice abuse Intestinal fistula Renal tubular failure Hypokalaemic periodic paralysis—intermittent weakness lasting <72h	Fluid loss without water replacement (e.g. diarrhoea, vomiting, burns) Diabetes insipidus—suspect if large urine volume Osmotic diuresis Primary aldosteronism: suspect if ↑ BP, ↓ K^+, alkalosis	Drugs—especially antidepressants (SSRIs most common) or diuretics (thiazides most common) Renal failure or nephrotic syndrome Gut—diarrhoea/vomiting; fistula; rectal villous adenoma; small bowel obstruction Endocrine—SIADH (➔ p. 341); severe hypothyroidism; Addison's disease; gluco-corticoid deficiency Cardiac failure Cirrhosis Cystic fibrosis (➔ p. 300) Heat exposure Water overload (e.g. polydipsia)

Postural (orthostatic) proteinuria 2–5% adolescents; rare >30y. Proteinuria disappears on early-morning sample. No long-term effects.

Urine ACR and PCR Check using a laboratory urine sample if persistent proteinuria on dipstick, or annually for all patients with DM or eGFR <60mL/min/1.73m^2. If initial result is abnormal and ACR<70mg/mmol, confirm with early morning sample. *Definitions*:

- Microalbuminuria ACR >2.5mg/mmol (♂) or >3.5mg/mmol (♀) or albumin concentration >20mg/L
- Proteinuria ACR ≥30mg/mmol or albumin concentration >200mg/L

❗ Use ACR in preference to PCR in most patients as more sensitive to low levels of proteinuria. Use PCR in preference to ACR if high levels of proteinuria, in children (<18y) and in pregnant women.

ACR and risk of adverse outcomes Figure 13.1, ➔ p. 411.

In people without diabetes Consider proteinuria to be clinically significant if ACR is ≥30mg/mmol (approximately equivalent to PCR ≥50mg/mmol, or a urinary protein excretion ≥0.5g/24h).

In people with diabetes Consider microalbuminuria to be clinically significant if ACR >2.5mg/mmol in ♂ or ACR >3.5mg/mmol in ♀.

Estimating renal function

Direct measure of glomerular filtration rate (GFR) using 24h plasma or urinary clearance is most accurate but is time-consuming and difficult in practice. Instead, other methods are often used to estimate GFR.

> Renal function ↓ with age (~1mL/min/y >40y). Always assume a degree of renal impairment in all patients >75y.

Urine ACR and PCR ➜ p. 409

Estimated glomerular filtration rate (eGFR) Method of calculation of renal function based on serum creatinine levels. Various equations exist but, in the UK, laboratories report eGFR using the Chronic Kidney Disease Epidemiology Collaboration (CKD-EPI) formula. This is adjusted for body surface area and uses age, sex (× 1.018 if ♀) and ethnicity (× 1.159 if of Afro-Caribbean origin) as variables.

Limitations of eGFR calculation Do not use if rapidly changing renal function or AKI. May be affected by some drugs (e.g. trimethoprim) and dependent on muscle mass and diet:

- Overestimates eGFR If elderly, low-protein diet, amputee or muscle-wasting disorder (e.g. myasthenia gravis, late-stage muscular dystrophy)
- Underestimates eGFR If high muscle mass (e.g. high-level sport or body builder), high protein diet (e.g. taking protein supplements), muscle breakdown (e.g. after heavy exercise, myositis, muscular dystrophy)

Interpretation of eGFR Table 13.2 and Figure 13.1

eGFR cystatin C Method of calculating eGFR using blood cystatin C levels instead of creatinine. Consider at initial diagnosis if:

- Standard eGFR (using creatinine) is 45–59mL/min/1.73m^2, sustained for ≥90d, and
- No proteinuria (ACR <3mg/mmol) or other marker of kidney disease

Under these circumstances, do not diagnose CKD if eGFR cystatin C >60mL/min/1.73m^2.

! Interpret eGFR cystatin C with caution if uncontrolled thyroid disease: falsely ↑ with hypothyroidism and ↓ with hyperthyroidism.

Cockcroft and Gault formula Box 13.1. Preferred method for estimating renal function if elderly (≥75y), high or low BMI (>40kg/m^2 or <18kg/m^2), or at extremes of muscle mass. Provides an estimate of creatinine clearance (CrCl).

Box 13.1 Formula for calculating CrCl

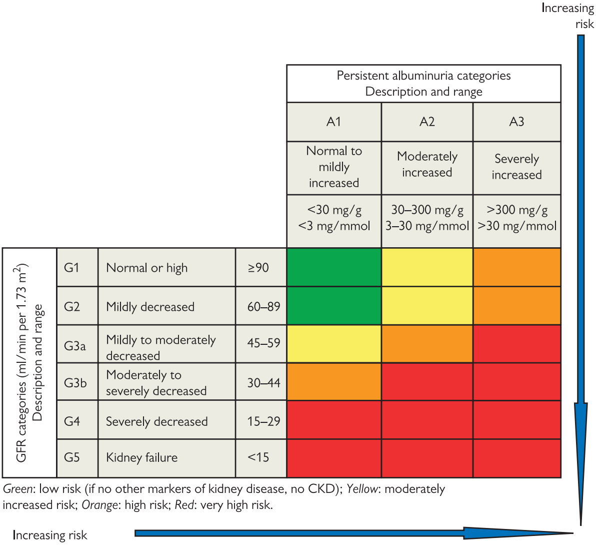
CrCl = [(140 − age in years)] × (weight in kg) ÷ serum Cr in micromol/L × 1.23 (♂) or 1.04 (♀)

! Use ideal body weight where fat may be a major contributor to body mass. Use actual body weight if <ideal body weight or high BMI is due to ↑ muscle bulk.

Ideal body weight in kg = 50 (♂) or 45 (♀) + 0.91 x (height in cm − 152.4)

Table 13.2 Interpretation of eGFR results

eGFR in mL/min/1.73m2			
>90	60–90	45–59	<45
Normal eGFR ❗ If serum Cr ↑ >20%, may indicate significant ↓ in renal function	Does not indicate CKD unless other markers of kidney damage are present, e.g. ACR ≥3mg/mmol, persistent microscopic haematuria, or structural abnormality, e.g. polycystic kidneys	May indicate CKD if proteinuria or other markers of kidney disease are present. If no proteinuria (ACR <3mg/mmol) and no other markers of kidney disease, consider checking eGFR cystatin C	On 2 readings >90d apart, indicates CKD (➔ p. 414)



Increasing risk
Increasing risk

				Persistent albuminuria categories Description and range		
				A1	A2	A3
				Normal to mildly increased	Moderately increased	Severely increased
				<30 mg/g <3 mg/mmol	30–300 mg/g 3–30 mg/mmol	>300 mg/g >30 mg/mmol
GFR categories (ml/min per 1.73 m^2) Description and range	G1	Normal or high	≥90	🟩	🟨	🟧
	G2	Mildly decreased	60–89	🟩	🟨	🟧
	G3a	Mildly to moderately decreased	45–59	🟨	🟧	🟥
	G3b	Moderately to severely decreased	30–44	🟧	🟥	🟥
	G4	Severely decreased	15–29	🟥	🟥	🟥
	G5	Kidney failure	<15	🟥	🟥	🟥

Green: low risk (if no other markers of kidney disease, no CKD); *Yellow*: moderately increased risk; *Orange*: high risk; *Red*: very high risk.

** For patients with eGFR ≥60mL/min/1.73m^2, if strong suspicion of CKD, check 24h urinary albumin/protein excretion*

Figure 13.1 Classification of CKD using eGFR and ACR categories

Reprinted from *Kidney International*, **85**, Levin A *et al*., Summary of KDIGO 2012 CKD Guideline: behind the scenes, need for guidance, and a framework for moving forward, 49–61. Copyright © 2013, with permission from the International Society of Nephrology.

Presentation of renal disease

Renal disease may present to the GP with:

- Haematuria (➔ p. 420)
- Proteinuria (➔ p. 408)
- ↓ renal function (➔ p. 410)
- Outflow tract obstruction (➔ p. 428)
- Hypertension (➔ p.218)
- UTI/pyelonephritis (➔ p. 422)
- Nephrotic syndrome
- Nephritic syndrome

Acute kidney injury (AKI)[N] ↓ renal function over hours/days ± oliguria/anuria. Investigate for AKI by checking serum Cr and comparing with baseline if acutely unwell and any of the following factors apply:

- Age >65y
- Heart failure
- Sepsis
- Past history of AKI
- Liver disease
- DM
- CKD (especially if eGFR is usually <60mL/min/1.73m^2)
- Hypovolaemia (e.g. diarrhoea, vomiting, diuretics, fever)
- Disability that may limit access to fluids (e.g. cognitive impairment, neurological condition)
- Potentially nephrotoxic drugs in the past week (e.g. NSAIDs, ACE inhibitor/ARB, diuretics, iodinated contrast agent)
- Oliguria (urine output <0.5mL/kg/h)
- History of, or condition predisposing to, urological obstruction

Diagnostic criteria for AKI

- ↑ in serum Cr of ≥26 micromol/L in <48h
- ≥50% ↑ in serum Cr known/presumed to have occurred in <7d
- ↓ in urine output to <0.5mL/kg/h for >6h in adults and >8h in children/young people
- ≥25% ↓ in eGFR in children/young people in <7d

Management If AKI is suspected, admit to hospital as an emergency.

Follow-up Monitor for development or progression of CKD for at least 2–3 years after AKI, even if Cr has returned to baseline.

Prevention Consider 'sick day rules'. If a patient is unwell with fevers, sweats/rigors, or vomiting/diarrhoea (unless minor), stop:

- ACE inhibitors and ARBs, e.g. losartan, candesartan, ramipril, lisinopril
- NSAIDs, e.g. ibuprofen, naproxen
- Diuretics, e.g. furosemide, bumetanide, bendroflumethiazide
- Metformin (↑ risk lactic acidosis when dehydrated)

Chronic kidney disease (CKD) ➔ p. 414

Nephrotic syndrome Proteinuria, hypoalbuminaemia, and oedema. Often associated with ↑ cholesterol. *Causes*:

- Minimal change glomerulonephritis (GN) (90% children, 30% adults)
- Membranous GN
- Focal segmental glomerulosclerosis
- Membranoproliferative GN
- DM
- Amyloid
- Neoplasia
- Endocarditis
- PAN
- SLE
- Sickle cell disease
- Malaria
- Drugs (penicillamine, gold)

Presentation Swelling of eyelids and face; ascites, peripheral oedema; urine froth due to protein. *Nephrotic crisis*: unwell with oedema, anorexia, vomiting, pleural effusions, and muscle wasting.

Investigation *Urine*: ACR (>220mg/mmol); microscopy for red cells and casts; *Blood*: U&E, creatinine, eGFR, albumin (<25g/L), cholesterol, FBC, ESR/CRP.

Management Refer all suspected cases of nephrotic syndrome to a renal physician. *Complications include*:

- Thromboembolism
- Hypercholesterolaemia
- Infection—especially pneumococcal—if persistent nephrotic syndrome, offer vaccination
- Hypovolaemia and renal failure
- Loss of specific proteins, e.g. transferrin (causes hypochromic anaemia which is iron resistant)

Nephritic syndrome Central feature is blood and protein in the urine from glomerular inflammation.

- Causes GN (may occur after throat, ear, or skin infection with group A β-haemolytic streptococci), vasculitis
- Features Oliguria, haematuria and proteinuria, fluid retention, ↑ BP, uraemia, and ↑ creatinine
- Management Refer suspected cases immediately to renal medicine
- Risks Hypertensive encephalopathy, pulmonary oedema, acute kidney injury
- Prognosis Excellent in children; in adults some proteinuria/urine sediment may persist. CKD is rare

Nephrocalcinosis Deposition of Ca^{2+} in the kidneys. *X-ray*: calcification. May cause symptoms of UTI or renal stones. *Cause*:

- Medullary (95%) Hyperparathyroidism, distal renal tubular acidosis, medullary sponge kidney, idiopathic calciuria, papillary necrosis, oxalosis
- Cortical Serious renal disease or chronic GN

Anaemia and renal disease 2° anaemia due to ↓ kidney erythropoietin production is universal among people with severe renal disease (G4/5). Exclude other causes. Recombinant erythropoietin is given if Hb <10.5g/dL.

Specific kidney diseases ➔ p. 416

Patient support and information

Kidney Patient Guide 🖱 www.kidneypatientguide.org.uk
The National Kidney Federation ☎ 0800 169 0936 🖱 www.kidney.org.uk

Chronic kidney disease

Slow ↓ renal function over months/years. Common (prevalence 8.5%); ↑ with age; ♀ > ♂. Usually asymptomatic. May be associated with ↑ BP. In later stages may cause nausea/vomiting, anorexia, lethargy, oedema, dyspnoea, ± itching.

Causes

- DM
- ↑ BP
- Urinary tract obstruction
- Chronic pyelonephritis
- Nephrotoxic drugs
- CVD
- ↑ Ca^{2+}
- Polycystic kidneys
- Glomerulonephritis
- Renovascular disease
- Interstitial nephritis
- SLE
- Amyloid
- Myeloma
- PAN
- AKI

Classification of CKD Figure 13.1 (➔ p. 411). Both ↑ ACR and ↓ eGFR are associated with ↑ risk of adverse outcomes (renal failure and CVD). In combination, risk is multiplied.

Testing for CKD^{N} Check eGFR + ACR (in most cases 1×/y) if ≥1 of:

- ↑ BP
- DM (all type 2; type 1 if present >5y)
- AKI (➔ p. 412)
- Haematuria (➔ p. 420)
- CVD (stroke, TIA, IHD, heart failure, peripheral vascular disease)
- Structural renal tract disease, recurrent renal calculi, or BPH
- Multisystem disease with potential kidney involvement, e.g. SLE
- FH of end-stage kidney disease or hereditary kidney disease
- If prescribed drugs known to be nephrotoxic, e.g. calcineurin inhibitors (e.g. ciclosporin, tacrolimus), lithium, NSAIDs

❗ Repeat test in <2wk if eGFR <60mL/min/1.73m^2 and no previous test.

Frequency of CKD monitoring^{N} Table 13.3. Tailor frequency of testing to the individual according to underlying cause of CKD, eGFR/ACR history, co-morbidities/general health status, intercurrent illness, and changes in drug treatment.

Investigation of haematuria ➔ p. 420

Refer for renal USS If:

- eGFR of <30 mL/min/1.73m^2 (G4/5) or accelerated progression of CKD: sustained ↓ in eGFR of ≥25% and a change in GFR category in <12mo, or sustained ↓ in eGFR of ≥15mL/min/1.73m^2 per year
- Symptoms of urinary tract obstruction
- Family history of polycystic kidney disease and aged >20y

Table 13.3 Frequency of CKD monitoring based on eGFR and ACR levels

eGFR category	ACR category		
	A1	A2	A3
G1	≤1×/y	1×/y	≥1×/y
G2	≤1×/y	1×/y	≥1×/y
G3a	1×/y	1×/y	2×/y
G3b	≤2×/y	2×/y	≥2×/y
G4	2×/y	2×/y	3×/y
G5	4×/y	≥4×/y	≥4×/y

Management of CKD Discuss causes of CKD, risks (progression of renal disease, ↑ BP, CVD, ↑ mortality) and management:

- Treat reversible causes if possible; stop/avoid nephrotoxic drugs (e.g. NSAIDs). Consider ↓ dose of other drugs as excretion/metabolism may be impaired. ❗ Stop metformin if eGFR <30mL/min/1.73m^2
- Annual review of CVD risk—check lipid profile; lifestyle advice (smoking, weight ↓, exercise, alcohol consumption)—➔ p. 219
- Treat ↑ BP (➔ p. 219)—target <140/90mmHg; if CKD + DM or CKD + ACR ≥70mg/mmol, aim for BP <130/80mmHg
- Offer ACE inhibitor/ARB if CKD + DM + ACR ≥3mg/mmol; CKD + ↑ BP + ACR ≥30mg/mmol; or CKD + ACR ≥70mg/mmol
- Offer atorvastatin 20mg nocte (↓ CVD events and death by 20%). ↑ dose if <40% ↓ in non-HDL cholesterol and eGFR ≥30mL/min/1.73m^2. If eGFR <30mL/min/1.73m^2, seek specialist advice
- Offer folic acid/vitamin B_{12} supplements if ↓ on laboratory testing/poor diet

Refer To renal physician if:

- eGFR <30mL/min/1.73m^2 *or* ACR ≥70mg/mmol (unless caused by DM and already treated) *or* ACR ≥30mg/mmol + haematuria (unless urgent referral for suspected cancer is indicated—➔ p. 420)
- Sustained ↓ in eGFR of ≥25% + change in GFR category or sustained ↓ in eGFR of ≥15mL/min/1.73m^2 in <12mo
- Poorly controlled BP despite ≥4 antihypertensive drugs
- Known/suspected rare/genetic causes of CKD or renal artery stenosis

If renal outflow obstruction Refer to urology.

Dialysis Starts if GFR <10–15% normal. Needed lifelong unless transplant becomes available. Refer to the patient's renal unit if any problems.

- **Haemodialysis** Blood flows opposite dialysis fluid and substances are cleared along a concentration gradient across a semi-permeable membrane. *Problems*: pulmonary oedema; infection (hepatitis, bacteria); U&E imbalance; BP ↑ or ↓; problems with vascular access; dialysis arthropathy (especially shoulders and wrists); aluminium toxicity; time
- **Continuous ambulatory peritoneal dialysis (CAPD)** Permanent catheter is inserted into the peritoneum via sc tunnel. Dialysis fluid is introduced and kept in the peritoneum; changed up to 5×/d at home. Patient is not tied to a dialysis machine. *Problems*: emergency referral—peritonitis, catheter blockage; other problems—weight ↑, poor DM control, pleural effusion, leakage

Renal transplantation Usually sited in an iliac fossa. 5y graft survival is ~88% for adults. Closer genetic matches → ↑ survival rates. *Problems*:

- Rejection
- Persistent ↑ BP
- Atherosclerosis (5× ↑ risk MI death) and ↑ cholesterol
- Renal artery stenosis at 3–9mo post-op
- Obstruction at ureteric anastomosis
- Ciclosporin-induced nephropathy
- Infection 2° to immunosuppression
- Malignancy from immunosuppressants

Further information

NICE (2014, updated 2015) Chronic kidney disease in adults. 🖱 www.nice.org.uk/guidance/cg182

NICE (2013) Acute kidney injury. 🖱 www.nice.org.uk/guidance/cg169

Specific kidney diseases

Interstitial nephritis Important cause of renal failure. Associated with inflammatory cell infiltration of the renal interstitium/tubules. *Causes*:

- Acute interstitial nephritis Idiosyncratic reaction to drugs (penicillin, NSAIDs, furosemide), or infection (*Staphylococcus* or *Streptococcus*)
- Chronic interstitial nephritis Idiopathic (most), drugs, sickle cell disease, analgesic nephropathy

Presentation and prognosis Presents with AKI/CKD, fever, arthralgia, eosinophilia. Patients with AKI have good prognosis. Those with CKD tend to gradual deterioration over time.

Diabetic nephropathy ➔ p. 325

Analgesic nephropathy Caused by prolonged heavy use of analgesics (including NSAIDs). Presents with an interstitial nephritis-like picture. Associated with ↑ incidence UTI. Carcinoma of the renal pelvis is a rare complication. Investigate promptly if the patient develops haematuria.

Glomerulonephritis Types and presentation—Table 13.4. Refer all suspected cases urgently to a renal physician. *Terminology*:

- Focal—some glomeruli affected
- Diffuse—all glomeruli affected
- Segmental—part of each glomerulus affected
- Global—all of each glomerulus affected

Chronic pyelonephritis Presents as CKD or one of its complications. Probably arises from UTIs, vesico-ureteric reflux, and consequent renal scarring in childhood (➔ p. 856). Refer to a renal physician.

Table 13.4 Types of glomerulonephritis

Type	Features
Minimal change	Most common in children. Presents with nephrotic syndrome
Membranous	30% adult nephrotic syndrome. Underlying malignancy in 10% of adults. 1 in 3 enter remission, 1 in 3 are proteinuric, 1 in 3 progress to end-stage renal disease (ESRD)
Focal segmental glomerulosclerosis	Proteinuria or nephrotic syndrome. May be associated with heroin misuse. >50% progress to CKD
Membrano-proliferative	50% present as nephrotic syndrome. Associations—endocarditis, C3 nephritic factor (autoantibody), hepatitis C, measles
Proliferative	Presents with nephritic syndrome. Classically seen 2wk after streptococcal infection. Prognosis is excellent
IgA disease (Berger's disease)	Causes recurrent haematuria in young men. A similar histological picture is seen in Henoch–Schönlein purpura (➔ p. 500). 30% progress to ESRD
Rapidly progressive/ crescentic	Presents with haematuria, oliguria, ↑ BP, acute kidney injury. Vigorous treatment may preserve renal function. *Causes*: anti-glomerular basement membrane disease (Goodpasture's disease), Wegener's granulomatosis, Henoch–Schönlein purpura

Haemolytic uraemic syndrome Most common cause of AKI in children. Usually follows gastroenteritis. Due to *E. coli* toxin. Have a high index of suspicion in any child with bloody diarrhoea. Occasionally occurs without diarrhoea. *Other features*:

- Dehydration
- Oliguria (though may be polyuria)
- Proteinuria/haematuria
- Haematological features—anaemia, thrombocytopenia ± purpura
- CNS symptoms—irritability, drowsiness, ataxia, coma
- ↑ BP (associated with non-diarrhoeal disease)

Management Admit for specialist care—often including dialysis. If associated with diarrhoeal illness, >80% make full recovery. Mortality is 1.8%. Poor prognostic indicators are age >5y at onset and dialysis for >2wk. Disease in the absence of diarrhoea has poorer prognosis.

Adult polycystic kidney disease Autosomal dominant disease (1 in 1000). Cysts develop in the kidney causing gradual ↓ in renal function. Common cause of CKD. Presents with haematuria, UTI, abdominal mass (30% have cysts in the liver/pancreas too), lumbar/abdominal pain, and/or ↑ BP. May be associated with mitral valve prolapse, and SAH/berry aneurysms. USS shows large kidneys with multiple cysts. Refer to a renal physician if CKD 3–5. Treat infections and ↑ BP. Check family members (though cysts may not be seen <30y). 45% progress to ESRD by 60y.

Medullary sponge kidney Developmental abnormality of the medullary pyramids of the kidney, characterized by dilatation of renal collecting tubules. ♂ > ♀. There may be a family history. Most are asymptomatic and the condition is an incidental finding. If symptomatic, presents with UTIs, renal stones, haematuria. Refer if symptomatic. Usually prognosis is very good and most require no treatment.

Renal vein thrombosis (RVT) *Causes*: nephrotic syndrome (15–20% develop RVT); membranous glomerulonephritis (30%); acute dehydration. Presentation varies from no symptoms to severe pain and loin tenderness. Suspect in at risk individuals if unexplained loss of renal function and RBCs in urine. Refer to a renal physician for further investigation.

Renal artery stenosis *Causes*: atheroma, fibromuscular hyperplasia (in the young). Presents with ↑ BP (may be severe or drug resistant); vascular disease elsewhere; abdominal bruit; ↑ Cr, ↓ eGFR and proteinuria. If bilateral or extensive, renal failure may be precipitated by dehydration, ↓ BP or drugs (ACE/ARB initiation; NSAIDs). Refer to a renal physician (if diagnosis is unsure) or vascular surgeon (if diagnosis is known).

Alport's syndrome X-linked or autosomal disease. Congenital sensorineural deafness, haematuria, proteinuria, and renal failure. Associated with lens abnormalities, platelet dysfunction, and ↑ BP. Causes ESRD by 3rd decade in ♂; ♀ rarely develop ESRD. Renal failure does not recur after transplantation.

Patient support and information

The National Kidney Federation ☎ 0800 169 0936 🖱 www.kidney.org.uk

Renal stones

12% of ♂ and 3% of ♀ will develop a renal stone at some point; peak age 20–50y. Symptoms are not dependent on size of the stone.

Risk factors

- Family history—↑ risk ×3. *Specific conditions*: X-linked nephrolithiasis, cystinuria, hyperoxaluria
- Anatomically abnormal kidneys—e.g. horseshoe kidney, medullary sponge kidney
- Metabolic disease—e.g. gout, hypercalcaemia/hypercalciuria, cystinuria, renal tubular acidosis or other acidosis (ileostomy, adenomatous polyp), oxaluria, aminoaciduria
- Dehydration
- Immobilization
- Chronic UTI

Drugs predisposing patients to stone formation Acetazolamide, allopurinol, aspirin, steroids, indinavir, nelfinavir, loop diuretics, probenecid, quinolones, sulfonamides, theophylline, thiazides, triamterene, antacids, calcium/vitamin D supplements, high-dose vitamin C.

Presentation Usually presents with pain ± nausea/vomiting. Location and type of pain gives clues about the site of the stone:

- *Loin pain*—kidney stone
- *Renal colic*—ureteric stone
- *Strangury*—bladder stone
- *Interruption of flow*—urethral stone

Renal colic

- **Symptoms** Severe pain with waves of ↑ severity. Usually starts abruptly as flank pain which then radiates around the abdomen to the groin as stone progresses down the ureter. May be referred to testis/tip of penis in man or labia majora in women
- **Signs** Patient is obviously in pain—usually unable to sit still and keeps shifting position to try to get comfortable (in contrast to peritonitis where patients tend to keep still). May be pale and sweaty. May be mild tenderness on deep abdominal palpation or loin tenderness, although often minimal signs. If fever suspect infection

Other presentations UTI, haematuria, retention, renal failure (rare).

Differential diagnosis Pyelonephritis; ruptured AAA; cholecystitis; pancreatitis; appendicitis; diverticulitis; obstruction; strangulated hernia; testicular torsion; pethidine addiction.

Immediate investigations Dipstick urine if possible. Absence of RBCs does not exclude renal colic but consider alternative diagnosis.

Immediate management Stones usually pass spontaneously. Give pain relief (diclofenac 75mg IM/100mg PR) ± antiemetic. Consider admission to hospital if:

- Fever
- Oliguria
- Poor intake of fluid
- Pregnant
- Lives alone
- Uncertain diagnosis
- Analgesia ineffective/short-lived
- Symptoms >24h

If not admitted Encourage ↑ fluid intake; sieve urine for stones. Monitor/review pain relief and for complications.

Further investigations Can wait until the next working day and include:

- **Blood** U&E, creatinine, eGFR, Ca^{2+}, PO_4^{3-}, alkaline phosphatase, uric acid, albumin
- **Urine** M,C&S; RBCs. Consider checking 'spot' test for urine cystine, and ACR, Ca^{2+}, PO_4^{3-}, uric acid, and sodium excretion
- **Radiology** X-ray of kidneys, ureters, and bladder—90% of renal stones are radio-opaque—only urate and xanthine stones are radio-translucent; renal tract USS

Follow-up 50% recur in 5–7y. Give general advice on prevention of stones (Table 13.5). If investigations show any loss of renal function, renal obstruction, or remaining stones—refer to urology. Dependent on composition of stones, give dietary advice/refer to dietician (Table 13.5).

Hyperoxaluria May be 1° (autosomal recessive condition) or 2° to gut resection/malabsorption or dietary excess of spinach or vitamin C. Take specialist advice on management. There are 2 types of 1° hyperoxaluria:

- **Type 1 hyperoxaluria** Calcium oxalate stones are widely distributed throughout the body. Presents as renal stones and nephrocalcinosis in children. 80% have chronic renal failure in <20y
- **Type 2 hyperoxaluria** More benign but less common—nephrocalcinosis but no chronic renal failure

Cystinuria Most common aminoaciduria. Usually presents with stones at age 10–30y. *Urine*: cystine ↑, ornithine ↑, arginine ↑, lysine ↑. Take specialist advice on management.

Hypercalcaemia ➜ p. 336

❗ Hypercalciuria may occur without hypercalcaemia and is found in ~80% of patients with calcium oxalate stones.

Table 13.5 Prevention of renal stones

Type of stone	Preventative measures
All types	↑ fluid intake (>2–2.5L/24h) especially in hot weather; ↓ weight if obese; ↓ animal protein and ↑ fruit/vegetables in diet; ↓ salt intake
Calcium oxalate	Urinary alkalinization with potassium citrate; avoid chocolate, tea, rhubarb and spinach, nuts, beans, beetroot; ↓ citrus fruits; bendroflumethiazide 2.5mg od may help if hypercalciuria; hyperoxaluria is treated with pyridoxine
Calcium phosphate	Low Ca^{2+} diet; avoid vitamin D supplements. Bendroflumethiazide 2.5mg od may help if hypercalciuria
Staghorn/triple phosphate (calcium, magnesium, and ammonium)	Associated with UTI due to *Proteus* spp. and urinary stasis, e.g. due to anatomical abnormality. Treat UTI with antibiotics
Urate	Avoid beer as has uricosuric effect; allopurinol; urinary alkalinization with potassium citrate (pH >6.5)
Cystine	Urinary alkalinization with potassium citrate

Haematuria, bladder and renal cancer

Haematuria^N Blood in the urine. Detected on urine dipstick. ≥1+ blood on urine dipstick is significant. *Causes*: Table 13.6.

- May be visible (frank) or non-visible (microscopic—up to 20% of the population). Non-visible haematuria (NVH) may be symptomatic (e.g. dysuria, ↑ frequency) or non-symptomatic (usually found by chance)
- Investigate *all* cases of haematuria: check BP; palpate the abdomen; send MSU for M,C&S
- Free Hb and myoglobin make urine test sticks +ve in absence of red cells. Urine discolouration can also result from beetroot ingestion, porphyria, bilirubinuria 2° to biliary obstruction or drugs, e.g. rifampicin
- If cause is identified (e.g. sample taken when menstruating, UTI)—repeat the check for blood in urine once treated/resolved

Refer urgently to urology To be seen in <2wk if:

- Child with unexplained haematuria; consider admission if unwell
- Age ≥45y with unexplained visible haematuria without UTI or visible haematuria that persists/recurs after successful treatment of UTI
- Age ≥60y with unexplained NVH + dysuria or ↑ WCC on blood test

Further primary care investigation If urgent referral not indicated:

- Check renal function: urine sample for ACR; blood for eGFR
- Consider PSA testing + DRE for ♂ with visible haematuria
- Refer for pelvic USS if ♀, age ≥55y, visible haematuria + unexplained vaginal discharge, ↓ Hb, ↑ platelets or ↑ blood glucose
- Refer for renal USS if visible or persistent NVH (on >1 urine sample)

Non-urgent referral

- Urology/gynaecology—age ≥60y + recurrent/persistent UTI
- Urology—any age + unexplained visible or symptomatic NVH, or age ≥40y and asymptomatic NVH
- Nephrology—eGFR <30mL/min/1.73m^2 or evidence of declining eGFR; significant proteinuria (ACR ≥30mg/mmol); age <40y with isolated haematuria (without proteinuria) + ↑ BP; visible haematuria coinciding with intercurrent infection (usually URTI)

❗ If ongoing persistent NVH without proteinuria/CKD, follow-up 1×/y with BP, urine dipstick for haematuria, ACR, and eGFR until resolves.

Table 13.6 Causes of haematuria

Infection	UTI, urethritis, prostatitis, TB, schistosomiasis, infective endocarditis
Tumour	Bladder, prostate, kidney or endometrial. Wilm's tumour (➔ p. 884)
Inflammation	GN, Henoch–Schönlein purpura, IgA nephropathy, Goodpasture's syndrome, polyarteritis, post-irradiation
Structural	Stones (renal, bladder, ureteric), cysts (simple cysts, polycystic renal disease), BPH, congenital vascular anomalies
Blood	Sickle cell disease, coagulation disorders
Trauma	Surgery/catheter/foreign body, sexual activity, abdominal/back injury
Drugs	Sulfonamides, cyclophosphamide, NSAIDs
Others	Menstruation, fabricated/induced illness

Sterile pyuria White cells in the urine without UTI. Repeat with clean catch MSU. If persists, refer to urology. *Causes*:

- Inadequately treated UTI
- Appendicitis
- Calculi
- Prostatitis
- Bladder tumour
- Renal TB
- Papillary necrosis
- UTI with failure to culture organism
- Interstitial nephritis or cystitis
- Polycystic kidney
- Chemical cystitis, e.g. due to radiotherapy

Bladder cancer *Incidence*: ~11,000 cases/y in the UK; ♂:♀ ≈5:2. Transitional cell carcinoma (TCC) is most common in the UK—squamous cell carcinoma (SCC) is most common worldwide. *Risk factors*: smoking (½ male cases are attributable to smoking); aromatic amine exposure (textile or rubber industries); stasis of urine; chronic UTI; schistosomiasis (SCC). Presents with haematuria. *Less commonly*: recurrent UTI, frequency, pelvic or loin pain, and/or bladder outflow obstruction.***Investigation and management*** MSU—excludes UTI and detects sterile pyuria; check urine dipstick for NVH. Refer to urology to be seen in <2wk. Treatment depends on stage at diagnosis—Table 13.7.

Table 13.7 Stage of bladder cancer, treatment, and prognosis

Stage	Description and treatment	Prognosis
T1 (80%)	Disease confined to mucosa/submucosa. Treated with transurethral resection of the tumour (TURBT) ± single intravesical chemotherapy	Very good—most die from other causes
T2	Invasion into connective tissue around the bladder. Treatment is with TURBT ± radiotherapy	60% survive 5y
T3	Invasion through muscle into the fat layer. Treated with radical cystectomy and/or radiotherapy	40–50% 5y survival
T4	Spread beyond the bladder. TURBT for local symptoms, palliative radio- ± chemotherapy	20–30% 5y survival

Investigation and management *Urine*: RBCs. *Blood*: ↑ PCV (2%), anaemia, hypercalcaemia. *Radiology*: USS, CXR. Refer to urology to be seen in <2wk. Treatment includes surgery where possible ± chemo-, radio-, and/or biological therapy. Overall 50% 5y survival.

Hypernephroma Clear cell adenocarcinoma of renal tubular epithelium. *Incidence*: 9300 cases/y in the UK. *Typical age*: 50y. ♂:♀ ≈1.5:1. Spread can be local or haematogenous (bone, liver, lung—causes cannon ball metastases seen on CXR). *Presentation*: haematuria, abdominal mass, loin pain, anaemia, left varicocele (♂) and, occasionally, night sweats.

Further information

NICE (2014, updated 2015) Chronic kidney disease in adults. 🖱 www.nice.org.uk/guidance/cg182

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Information and support for patients

Cancer Research UK ☎ 0808 800 4040 🖱 www.cancerresearchuk.org
Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk

Urinary tract infection

Urinary tract infection (UTI) is one of the most common conditions seen in general practice accounting for up to 6% of consultations (1 case/average surgery). ♀ > ♂. 20% of women at any time have asymptomatic bacteriuria and 20–40% of women will have a UTI in their lifetime.

Infecting organisms *E. coli* (>70%), *Proteus* spp., *Pseudomonas* spp., streptococci, staphylococci.

Risk factors

- Prior infection
- DM
- Stones
- Pregnancy
- Dehydration
- GU instrumentation
- Catheterization
- ↓ oestrogen (menopause)
- Sexual intercourse
- GU malformation
- Urinary stasis (e.g. obstruction)
- Delayed micturition (e.g. on long journeys)

Presentation of UTI

- **Cystitis** Frequency, dysuria, urgency, strangury, low abdominal pain, incontinence of urine, acute retention of urine, cloudy or offensive urine, and/or haematuria
- **Pyelonephritis** Loin pain, fever, rigors, malaise, vomiting, and/or haematuria

Dysuria and urgency Painful micturition resulting from urethral or bladder inflammation. *Causes*: UTI, urethral syndrome, inflammation (e.g. interstitial cystitis, radiation-induced cystitis), intravesical lesion (tumour, stone), atrophy (menopause).

Frequency Passage of urine more often than usual. *Causes*:

- UTI
- Urethral syndrome
- Detrusor instability
- Inflammation (e.g. interstitial cystitis)
- Fibrosis (e.g. post-radiotherapy)
- Atrophy (menopause)
- Neurogenic bladder (e.g. MS)
- External pressure (e.g. pregnancy, fibroids)
- Bladder tumour or stone
- Enlarged prostate
- Drugs (e.g. diuretics)
- DM
- Excessive fluid intake
- Habit

Initial investigation If uncomplicated UTI in an otherwise healthy ♀, test urine with a leucocyte and nitrite dipstick. If +ve treat for UTI. *Reasons to send MSU for M,C &S*:

- Unresolved infection after antibiotics
- Recurrent UTI
- Uncatheterized ♂ with UTI
- Catheterized ♂ or ♀ with symptomatic UTI
- Impaired renal function
- Child—➔ p. 856
- Pregnant ♀—➔ p. 798
- Suspected pyelonephritis
- Haematuria—microscopic or macroscopic—always investigate further—➔ p. 420
- Abnormal renal tract

❗ Take MSU prior to starting antibiotics—send to the laboratory fresh.

Further investigation Consider further investigation with blood tests (eGFR ± PSA if >40y and ♂) and/or radiology (e.g. renal tract USS) if:

- UTI in a ♂
- UTI in a child (➔ p. 856)
- Recurrent UTI in a ♀
- Pyelonephritis
- Unusual infecting organism
- Sterile pyuria (➔ p. 421)
- Unclear diagnosis (e.g. persisting symptoms but negative MSU)

Management

- Catheterized patients—➔ p. 427
- Pregnant ♀—➔ p. 798
- Children—➔ p. 856

All other patients[N] Symptomatic relief with paracetamol ± NSAIDs. ↑ fluid intake. Prescribe oral antibiotics, e.g. nitrofurantoin MR 100mg bd or trimethoprim 200mg bd (consider local guidelines/resistance):

- 3d course for ♀ with uncomplicated UTI—consider delayed prescription if mild symptoms
- 7–10d course for ♂ or if GU malformation, immunosuppression, relapse (same organism), or recurrent UTI (different organism)

Use a 7d course of a quinolone (e.g. ciprofloxacin 250–500mg bd) for pyelonephritis. Refer to urology if abnormalities are detected on further investigation or unable to resolve symptoms. Admission is rarely needed.

Prevention of recurrent cystitis Reinfection after successful treatment of infection (90%) or relapse after inadequate treatment.

- General advice Advise to urinate frequently; ↑ fluid intake; double void (i.e. go again after 5–10 min) and void after intercourse
- Prophylactic antibiotics Consider prescribing either postcoitally (e.g. nitrofurantoin MR 100mg stat) or continuously (trimethoprim 100mg or nitrofurantoin 50mg nocte)
- Men with BPH Finasteride/dutasteride and/or doxazosin ↓ UTI
- HRT Topical oestrogen ↓ recurrent UTI in ♀ of all ages
- Vaccines Results of large-scale trials are awaited

Prostatitis ➔ p. 430

Chronic pyelonephritis ➔ p. 416

Urethral syndrome Symptoms of cystitis with −ve MSU. Unknown cause. Associated with cold, stress, nylon underwear, COC, and intercourse. Advise fluids ++ and to wear cotton underwear. Consider changing/stopping COC, or trying topical oestrogen if postmenopausal. Tetracyclines (e.g. doxycycline 100mg bd for 14d) or azithromycin (500mg od for 6d) are helpful in some. If not settling, refer to urology.

Interstitial cystitis Predominantly middle-aged ♀. Can cause fibrosis of the bladder wall. *Main symptoms*: frequency, urgency, and pelvic/suprapubic pain especially when the bladder is full. Often misdiagnosed as recurrent UTI. *MSU*: no bacteriuria. Refer to urology for confirmation. There is no satisfactory treatment although antispasmodics, amitriptyline and bladder stretching under GA may help some patients.

Further information

NICE Antimicrobial prescribing:

- UTI (lower) (2018). 🖱 www.nice.org.uk/guidance/ng109
- UTI (recurrent) (2018). 🖱 www.nice.org.uk/guidance/ng112
- Acute pyelonephritis (2018). 🖱 www.nice.org.uk/guidance/ng111

Incontinence of urine

Involuntary loss of urine which is demonstrable and a social or hygienic problem. 1 in 3 with incontinence consult at outset, 1 in 3 consult later, 1 in 3 suffer in silence. Opportunistic questioning can identify sufferers.

History

- Frequency of complaint
- Volume passed
- Degree of incapacity
- Whether occurs with standing/coughing/sneezing
- Urgency/dysuria/frequency of micturition
- Past obstetric and medical history
- Medication
- Mobility and accessibility of toilets

Examination

- **Abdominal including DRE**—enlarged bladder, masses, loaded colon, faecal impaction, anal tone
- **Pelvic**—prolapse, atrophy, neurological deficit, retention of urine, and pelvic masses

Investigation Intake/output diary (at least 3d including working and leisure days)—evaluates problem and benchmark for progress (record drinks and passage of urine); Urine—RBCs, M,C&S; consider blood for U&E, eGFR FBC, FBG/HbA1c if renal impairment/DM is suspected.

Drugs that exacerbate/cause incontinence Diuretics, antihistamines, anxiolytics, α-blockers, sedatives and hypnotics, anticholinergic drugs, TCAs.

GP management ❗ 30% have a mixed pattern. Treat according to dominant symptom. Try general measures before referring to urology/gynaecology or for further investigations.

General measures

- Manipulate fluid intake: amount, type (avoid tea, coffee, alcohol), timing
- Promote weight ↓
- Alter medication, e.g. timing of diuretics
- Treat UTI and chronic respiratory conditions
- Avoid constipation
- Consider HRT (topical or systemic) for oestrogen deficiency
- Consider scheduled voiding if cognitive deficit
- Referral—Table 13.8

Aids and appliances for incontinence ➜ p. 426

Nocturnal enuresis in children ➜ p. 893

Stress incontinence

- **Symptoms** Small losses of urine without warning throughout the day related to coughing/exercise
- **Causes** Prostatectomy; childbirth; deterioration of pelvic floor muscles/nerves
- **Treatment** Pelvic floor exercises (➜ p. 821) continued >3mo help 60% (taught by physiotherapists/continence advisors; leaflets available)—may be assisted by vaginal cones and/or electrical stimulation. Mechanical devices (e.g. Contrelle Activguard®, FemSoft®) may help

Table 13.8 Referral for incontinence problems

Specialist continence advisor/DN	• Advice on aids or appliances • Advice on primary care management • Patient support
Urodynamic studies	• If type of incontinence is uncertain • Atypical features of incontinence • After unsuccessful surgery • If a neurological problem is suspected
Gynaecology or urology opinion	• GP management has failed • Severe symptoms and/or pain • Recurrent UTI • Concomitant gynaecological problems (e.g. prolapse) • Concomitant urological problems (e.g. chronic retention, prostate abnormality on rectal examination) • Failed incontinence surgery • Pelvic radiotherapy • Vesico-vaginal fistula • Haematuria—➔ p. 420

Urge incontinence (overactive bladder syndrome) Detrusor instability or hyperreflexia cause the bladder to contract unintentionally.

- **Symptoms** Frequency, overwhelming desire to void (often precipitated by stressful event), large loss, nocturia
- **Causes** Idiopathic, neurological problems (stroke, MS, DM, spinal cord injury, dementia, PD), local irritation (bladder stones, bladder cancer, infection), obstruction (BPH), surgery (TURP)

Treatment Bladder training—resist the urge to pass urine for ↑ periods. Start with an achievable interval based on diary evidence and ↑ slowly—continue for >6wk. If bladder training is ineffective, try oxybutynin first line[N] (alternatives: solifenacin, tolterodine, trospium, duloxetine). Spontaneously remits/relapses; reassess every 3–4 mo.

Overflow Constant dribbling loss day and night. *Causes*: BPH, prostate cancer, urethral stricture, faecal impaction, neurological (LMN lesions), side effect of medication. Treatment is aimed at relieving the obstruction (➔ p. 428).

Urinary fistula Communication between bladder and the outside—normally through the vagina. Results in constant dribbling loss day and night. Refer to gynaecology/urology. *Causes*: congenital, malignancy, complication of surgery.

Functional incontinence No urological problem. Caused by other factors, e.g. inaccessible toilets/immobility, behavioural problems, cognitive deficit. Treat the cause.

Further information

European Association of Urology (2019) Urinary incontinence. 🖱 http://uroweb.org/guideline/urinary-incontinence/

NICE (2019) Urinary incontinence and pelvic organ prolapse in women: management. 🖱 www.nice.org.uk/guidance/ng123

Patient information and support

Bladder and Bowel Foundation ☎ 01926 357220 🖱 www.bladderandbowel.org

Aids and appliances for incontinence

Pads Many different types. DNs or continence advisors are best aware of those available via the NHS locally. They are not available on FP10 and supplied by local NHS Trusts on a 'daily allowance' basis. This varies across the country.

Bed covers Absorb 1–4L of urine. Good laundry facilities are needed. If left wet can cause skin breakdown. Available via NHS Trusts.

External catheters or sheaths Can be prescribed on NHS prescription. Approved appliances are listed in part IXB of the UK Drug Tariff. Used for men who have intractable incontinence and who are highly physically dependent, do not have urine retention, and do not require an internal catheter. Used in association with a drainage bag. Assessment and fitting by a DN or continence adviser is essential.

May be non-adhesive, self-adhesive, or attached with adhesive strips. Adhesive sheaths can last several days but daily changing is recommended. Replace non-adhesive sheaths 2–3×/d (some are reusable).

Problems Include ↑ susceptibility to UTI, sores on penis, and skin irritation due to the adhesive.

Catheters Can be prescribed on NHS prescription. Approved appliances are listed in part IXA of the UK Drug Tariff.

Indwelling catheters Only use catheters in patients who have:
- Urinary retention or neurogenic bladder dysfunction
- Severe pressure sores
- Inoperable obstructions that prevent the bladder emptying
- Terminal illness
- Housebound without adequate carer support

Types Only long-term Foley catheters are suitable for use in primary care. They last 3–12wk.

Catheter size Unless specified a 12 or 14Ch catheter is supplied. Use the smallest diameter of catheter that drains urine effectively. Catheters >16Ch are more likely to cause bypassing of urine around the catheter and urethral strictures.

Catheter length Men require longer catheters than women. Specify 'male' or 'female' on the prescription.

Catheter balloon 10mL balloons are supplied unless specified otherwise. Pre-filled catheters contain sterile water which inflates the retaining balloon with water. They are more expensive but quicker to insert and there are no costs for syringes or sterile water.

Insertion ➔ p. 429

Drainage Usually attached to a leg bag, although catheter valves are also available allowing the patient to use his/her bladder as a urine reservoir. The valve must be released every 3–4h to drain out the urine

Common problems

- **Leakage** Check no constipation, check catheter not blocked, try smaller gauge catheter
- **Infection** 90% develop bacteriuria <4wk after insertion. Always confirm suspected UTI with MSU—only treat if symptomatic or *Proteus* species grown. May prove difficult to eliminate. No good evidence bladder instillations help
- **Encrustation** (50%) Deposition of minerals and other materials from the urine onto the catheter. Worse if there is infection with *Proteus* species. May cause catheter blockage or pain changing the catheter. Check pH of urine regularly in patients with problems. Citric acid patency solutions may help if pH >7.4 or a daily dose of vitamin C
- **Inflammation** Results from physical presence of a catheter in the urethra. Exacerbated by encrustation and infection. There is no easy solution—try a different brand catheter (e.g. hydrogel catheter rather than silicone)
- **Blockage** Change catheter. The interval of routine changes should be altered if there is regular blockage towards the end of the life of a catheter

Intermittent self-catheterization Patient inserts a catheter into his/her bladder 4–5×/d to drain urine. ↓ problems of infection and blockage. Useful for neurological bladder dysfunction. *Types*:

- Reusable silver or stainless steel
- Reusable PVC—washed and reused for 1 wk. Usually supply 5/mo
- Single use—need 125–150/mo. Expensive. Only use on consultant advice

Collecting bags Can be prescribed on NHS prescription. Approved appliances are listed in part IXB of the UK Drug Tariff.

- **Leg bags** Drainable bags last 5–7d. Usually 500/750mL. Larger capacity bags are too heavy for mobile patients. A variety of attachment systems are available on prescription. Long tubes are needed to wear a bag on the calf
- **Night drainage bags** Connect to night bag attachment of day bags. Single use, disposable non-draining bags are recommended. Bag hangers are not available on NHS prescription

Enuresis alarms Table 23.13, ➔ p. 893

Further information

NHSBSA Electronic drug tariff. 🖱 www.nhsbsa.nhs.uk/prescriptions
NICE (2015, updated 2017) Urinary tract infection (catheter-associated). 🖱 www.nice.org.uk/guidance/ng113

Patient advice and support

Bladder and Bowel Foundation ☎ 01926 357220 🖱 www.bladderandbowel.org

Urinary tract obstruction

Causes of obstruction Figure 13.2. Obstruction may be unilateral (kidney, pelvi-ureteric junction, or ureter), or bilateral (bladder, urethra, prostate). Unilateral obstruction may present late if the other kidney remains functioning. Suspect if loin ache worsened by drinking. Confirm with USS and refer to urology. Obstructing lesions may be in the lumen, e.g. stones; in the wall, e.g. tumours; or impinging from outside, e.g. retroperitoneal fibrosis.

Acute retention of urine Sudden inability to pass urine → lower abdominal discomfort with inability to keep still. Differentiate from AKI. ♂ > ♀. *Risk factors*: age >70y; symptoms of prostatism/poor urinary stream.

Causes Prostatic obstruction (82%); constipation; alcohol; drugs (anticholinergics, diuretics); UTI; operation (e.g. hernia repair). *Rarer causes*: urethral stricture; clot retention; spinal cord compression; bladder stone.

Examination Abdomen—palpable bladder; DRE—enlarged ± irregular prostate; perineal sensation to exclude neurological cause.

Investigation MSU to exclude infection. Blood for U&E, Cr, and eGFR. *Only* investigate if catheterizing in the community.

Management Catheterize (record initial volume drained) or refer to urology for catheterization—local policies vary. Treat infection. Refer to DN for instruction on management of the catheter. Refer to urology for further assessment and treatment.

Chronic retention of urine Insidious onset. *Causes*: benign prostatic hypertrophy; pelvic malignancy; CNS disease. May present as:

- Nocturnal enuresis
- Overflow incontinence
- Acute on chronic retention
- Lower abdominal mass
- UTI
- Renal failure

Examination and investigation As for acute retention. Bladder is enlarged (may contain >1.5L) but usually non-tender.

Management Refer to urology for further assessment and treatment. Refer urgently or acutely if pain, UTI, or renal failure (eGFR <60mL/min/1.73m^3). *Do not* catheterize in the community.

Retroperitoneal fibrosis Ureters become embedded in dense fibrous plaques in the retroperitoneal space. *Associations*:

- Drugs, e.g. methysergide
- Carcinoma
- Crohn's disease
- Connective tissue disease
- Raynaud's syndrome
- Fibrotic diseases, e.g. alveolitis

Presentation and management Typically middle-aged men presenting with fever, malaise, sweating, leg oedema, ↑ BP, palpable mass, and acute/chronic renal failure. Refer for specialist care. Options include steroids and nephrostomies.

Horseshoe kidney Congenital abnormality. Kidneys are fused in the midline to form a horseshoe-shaped mass. The kidney may function normally or may present with obstructive nephropathy or UTIs.

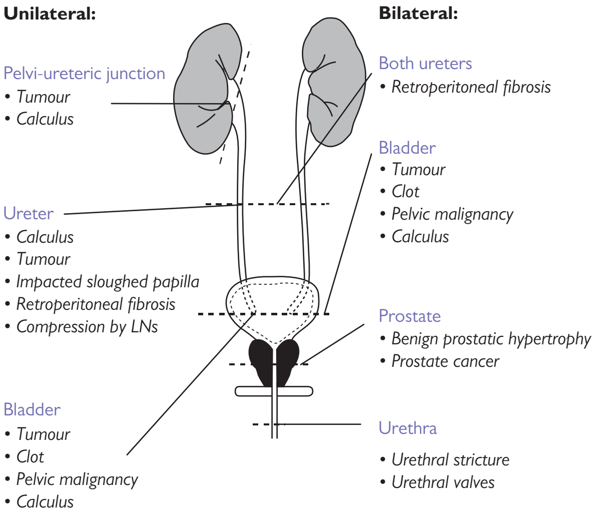

Unilateral:
Pelvi-ureteric junction
• Tumour
• Calculus
Ureter
• Calculus
• Tumour
• Impacted sloughed papilla
• Retroperitoneal fibrosis
• Compression by LNs
Bladder
• Tumour
• Clot
• Pelvic malignancy
• Calculus
Bilateral:
Both ureters
• Retroperitoneal fibrosis
Bladder
• Tumour
• Clot
• Pelvic malignancy
• Calculus
Prostate
• Benign prostatic hypertrophy
• Prostate cancer
Urethra
• Urethral stricture
• Urethral valves

Figure 13.2 Causes of urinary tract obstruction

Passing a urethral catheter ❗ Technique learned through supervised experience. Only attempt alone if you are competent to do so.

Prepare the equipment needed

- Sterile rubber gloves, plastic sheet to prevent spills, paper sheet to provide sterile field, cleansing materials—cotton swabs, cleansing fluid
- Local anaesthetic/lubricating gel, e.g. 1% lidocaine + 0.25% chlorhexidine
- Catheter—usually 12Ch or 14Ch—ensure the catheter is long if catheterizing a man (➔ p. 426)—and if the catheter is not pre-filled—sterile syringe + 10mL of sterile water
- Kidney dish/other receptacle to catch the urine before connecting the drainage bag; drainage tube and bag

Inserting the catheter

- Ensure the patient is comfortable; protect against spills with a plastic sheet; cover area with a sterile paper sheet
- Ensure strict aseptic technique. Cleanse the penis/vulva and squeeze lubricant/local anaesthetic gel into the urethra—allow it to work
- Gently but firmly, push the catheter into the urethra. Ensure the end of the catheter is over the receptacle. When the catheter enters the bladder, urine flows into the receptacle. Inflate the balloon with sterile water (if needed) once the catheter is inside the bladder. Connect the catheter to the collecting tube and bag
- If male, pull the foreskin over the glans again to prevent paraphimosis

❗ If you are unable to pass a catheter, refer to urology.

Benign prostatic hypertrophy

10–30% of men in their early 70s have symptomatic benign prostatic hypertrophy (BPH). There is no relation between size of the prostate and symptoms. *Assessment*—Table 13.9.

Symptoms of prostatism

- Obstructive ↓ and intermittent urinary stream, double micturition, hesitancy, terminal dribbling, feeling of incomplete emptying, and straining to void. *Differential diagnosis*: prostatic enlargement, strictures, tumours, urethral valves, bladder neck contracture
- Irritative (due to detrusor muscle hypertrophy)—urinary frequency, urgency, dysuria and nocturia. *Differential diagnosis*: enlarged prostate, UTI, polydipsia, detrusor instability, hypercalcaemia, uraemia

Complications 10% at presentation

- Recurrent UTI
- Bladder stones
- Haematuria
- Chronic obstruction—➔ p. 428
- Overflow incontinence—➔ p. 425
- Obstructive nephropathy
- Acute retention of urine ± prior obstructive symptoms—➔ p. 428

GP management Symptoms can improve spontaneously but overall progress slowly. 1–2%/y develop urinary retention. *Options*:

Watchful waiting Patients with mild to moderate symptoms at presentation with no complications of BPH and who are not severely troubled by their symptoms. Self-help includes: ↓ evening fluid intake, ↓ caffeine intake, bladder retraining and prevention of constipation.

Drug therapy Those with mild/moderate symptoms who are troubled by their symptoms. *Consider*:

- α-adrenoceptor agonists, e.g. prazosin, doxazosin—watch for postural hypotension. ↓ symptomatic worsening
- 5α-reductase inhibitors, e.g. finasteride—best for patients with bulky prostates—takes up to 6mo to work. ↓ risk of urinary retention
- Combination therapy—α-adrenoceptor agonist and 5α-reductase inhibitor ↓ progression by 66%—more than either agent alone

Referral to a urologist *E* = Emergency admission; *U* = Urgent; *S* = Soon; *R* = Routine

- Complicated BPH (e.g. acute retention)—*E/U*
- Nodular/firm prostate on DRE—*U*
- ↑ PSA (➔ p. 433)—*U*
- Severe symptoms—*S*
- Failure to respond to drug therapy after 3–12mo (α-blocker) or 6–12 mo (5α-reductase inhibitor)—*R*

Bacterial prostatitis Consider in men presenting with suspected UTI. *Other features*: fever; arthralgia/myalgia; low back, perineal, penile, ± rectal pain. DRE reveals swollen, tender prostate. If suspected, check MSU and treat with paracetamol/ibuprofen ± codeine and 4wk course of oral antibiotic which penetrates prostatic tissue, e.g. ciprofloxacin 500mg bd, ofloxacin 200mg bd. Refer for specialist advice if not settling. *Complications include*: acute retention of urine, chronic bacterial prostatitis, and prostate abscess.

Table 13.9 Assessment of BPH

Assessment	Comments
History	● General well-being ● Obstructive symptoms ● Irritative symptoms ● Haematuria ● Pain ● Polyuria and polydipsia ● Neurological symptoms ● Past history of urological instrumentation or STIs
Frequency–volume chart	Assess pattern and type of fluid consumption (e.g. alcohol/caffeine at night ↑ nocturia)
Symptom score (IPSS—➔ p. 432)	Objectively grade symptoms giving measure of severity. IPSS scores: 0–7 mild; 8–19 moderate; 20–35 severe A general quality of life measurement can be used to assess impact of symptoms
Abdominal examination	Look for distended bladder, palpable kidneys. Examine external genitalia
Digital rectal examination	Anal tone, size, shape, and consistency of prostate (normal prostate—size of a chestnut with smooth, rubbery consistency)
Serum urea, creatinine, and eGFR	Renal function assessment
MSU	Dipstick for blood and glucose. M,C&S
Ultrasound measurement of post-micturition residual	
Maximum voiding flow rate[a]	<15mL/s for voided volume. >100mL is abnormal
Serum PSA	High values can indicate prostate cancer (➔ p. 433)

[a] May be available through open-access prostate assessment clinics.

Chronic prostatitis (chronic pelvic pain syndrome) 2–14% lifetime prevalence. Cause is unknown. Presents with >3mo history of:

- Urological pain—lower abdomen, pelvis/perineum, penis (especially tip ± on ejaculation), testicles, rectum, low back ±
- Irritative/obstructive symptoms and/or ejaculatory disturbance

Diagnosis is based on history with exclusion of other causes. Suitable investigations include DRE, MSU, urine cytology, sexually transmitted infection screen, PSA, ± urodynamic studies. Treatment is difficult—provide information and support; try α-blockers (e.g. doxazosin 4mg od for 6mo). Spontaneous improvement/remission often occurs.

Further information

Rees J, et al. (2015) Diagnosis and treatment of chronic bacterial prostatitis and chronic prostatitis/chronic pelvic pain syndrome: a consensus guideline. *BJU Int* **116**:509–25. 🖱 https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5008168/pdf/BJU-116-509.pdf

Patient support

The Urology Foundation 🖱 https://www.theurologyfoundation.org/urologyhealth/prostate/prostatitis

The International Prostate Symptom Score

	Not at all	Less than 1 time in 5	Less than half the time	About half the time	More than half the time	Almost always	Your score
Over the past month, how often have you had a sensation of not emptying your bladder completely after you finish urinating?	0	1	2	3	4	5	
Over the past month, how often have you had to urinate again <2h hours after you finished urinating?	0	1	2	3	4	5	
Over the past month, how often have you stopped and started several times when you urinated?	0	1	2	3	4	5	
Over the past month, how often have you found it difficult to postpone urinating?	0	1	2	3	4	5	
Over the past month, how often have you had a weak urinary stream?	0	1	2	3	4	5	
Over the past month, how often have you had to push or strain to begin urinating?	0	1	2	3	4	5	
Over the past month, typically from the time you went to bed to the time you got up in the morning, how many times did you get up to urinate?	0	1	2	3	4	5+	
Total IPSS score							

	Delighted	Pleased	Mostly satisfied	Equally satisfied/ dissatisfied	Mostly dissatisfied	Unhappy	Terrible
If you were to live the rest of your life with your urinary condition the way it is now, how would you feel about it?	0	1	2	3	4	5	6

0–7 = mildly symptomatic; 20–35 = severely symptomatic; 8–19 = moderately symptomatic.

The International Prostate Symptom Score is reproduced with permission from the American Urological Association.

Prostate-specific antigen (PSA) testing There is no prostate screening programme in the UK but men can request a PSA test. The Government has introduced a PSA Informed Choice Programme. Warn patients about the poor specificity of the test before performing the test and provide information about the pros and cons of testing.

In addition, PSA is routinely measured in men with urological symptoms. Abnormal PSA is a common reason for referral to an urologist. Its sensitivity and specificity are poor.

Pros and cons of PSA testing

Benefits of PSA testing	Downside of PSA testing
• It may provide reassurance if the test result is normal • It may find cancer before symptoms develop and at an early stage when treatments could be beneficial • If treatment is successful, the consequences of more advanced cancer are avoided	• It can miss cancer, and provide false reassurance • It may lead to unnecessary anxiety and medical tests when no cancer is present • It might detect slow-growing cancer that may never cause any symptoms or shortened lifespan • The main treatments of prostate cancer have significant side effects, and there is no certainty that treatment will be successful

Reasons for ↑ PSA

- Prostate cancer
- Benign prostatic hypertrophy
- Acute or chronic prostatitis
- Physical exercise
- Acute urinary retention
- Prostate instrumentation (includes prostate biopsy and urinary catheterization)
- Old age

❗ PSA may be normal when early prostate cancer is present.

Performing a PSA test Digital rectal examination may cause a transient ↑ in PSA levels (💣), so do the PSA test before doing a digital rectal examination. If that is not possible, delay the test for 1wk after the examination. Exclude urinary infection before PSA testing. Do NOT do a PSA test if the man has:

- A proven UTI—treat the UTI and postpone the PSA test for ≥1mo
- Ejaculated within the previous 48h
- Exercised vigorously in the previous 48h
- Had a prostate biopsy <6wk ago

PSA cut offs that should prompt referral

Age (y)	Refer to urology if PSA (ng/mL)
50–59	≥3.0
60–69	≥4.0
≥70	>5.0

❗ Finasteride and dutasteride ↓ PSA by ~50%.

Prostate cancer

Prostate cancer is the 6th most common cancer worldwide. It is the most common cancer affecting men making up 26% of all male cancer diagnoses and ~10,000 men/y die from the disease in the UK. 1 in 6 men have clinical prostate cancer in their lifetimes and the incidence is rising.

Classification

Non-metastatic prostate cancer Can be divided into:

- Clinically localized disease—cancer thought after clinical examination to be confined to the prostate gland
- Locally advanced disease—cancer that has spread outside the capsule of the prostate gland but has not yet spread to other organs

Metastatic prostate cancer Cancer that has spread outside the prostate gland to local, regional, or systemic LNs, seminal vesicles, or other body organs (e.g. bone, liver, brain).

Risk factors

- **Age** Uncommon <50y; 85% are diagnosed aged >65y
- **Genetic** ↑ incidence if first-degree relative affected
- **Racial** Incidence varies according to location in the world and ethnic group. Highest rates are in men of black ethnic group in the USA—lowest in Chinese men
- **Dietary** Links are proposed between prostate cancer and low intake of fruit (particularly tomatoes) and high intake of fat, meat and Ca^{2+}

Screening There is currently no screening programme in the UK. *Problems with screening*:

- Incidental postmortem evidence of prostate cancer is high (≈75% men >75y), very few become clinically evident, so many more men would be found with prostate cancer by screening than would die or have symptoms from it
- Natural history of prostate cancer is not understood—there is no means to detect which 'early' cancers become more widespread
- Inadequate screening tests
- It is not clear if early treatment enhances life expectancy
- Peak incidence of morbidity and mortality is in old age (75–79y) so potential years of life saved by screening are small

Screening tests

- **Prostate-specific antigen (PSA)** ➔ p. 433
- **Digital rectal examination (DRE)** Operator-dependent, fails to detect early prostate cancers and lacks specificity. Annual screening in the USA and Germany has not ↓ mortality
- **Transrectal ultrasound (TRUS)** Too expensive

The most effective screening regime involves rectal examination and PSA testing followed by TRUS for suspicious lesions. Optimal screening interval is unknown but serial screening does ↑ detection.

Symptoms and signs

Early cancer Symptomless. Usually detected following an incidental finding of ↑ PSA. Hard nodule sometimes felt in prostate on DRE.

Local disease

- Prostatism
- Urinary retention
- Haematuria
- Lower extremity oedema
- On rectal examination, the prostate is hard, non-tender, and sulci lose definition

Metastatic disease

- Malaise
- Weight loss
- Bone pain
- Pathological fractures
- Spinal cord compression
- Ureteric obstruction may cause renal failure
- Signs depend on site of metastases

Investigation[N] A digital rectal examination and a PSA test (after counselling) are recommended for patients with any of the following unexplained symptoms:

- Erectile dysfunction
- Haematuria
- Lower back pain
- Any lower urinary tract symptoms
- Weight loss, especially in the elderly
- Bone pain

❗ Exclude UTI before PSA testing and postpone digital rectal examination until after the PSA test is done.

Urgent referral[N]

- Rectal examination—hard, irregular prostate typical of prostate cancer. PSA result should accompany the referral
- PSA levels are greater than the age-specific reference range

❗ Consider discussion with specialist and patient ± carer before referral for very elderly patients and those compromised by other co-morbidities.

❗ Referral is not needed if the prostate is simply enlarged and the PSA is in the age-specific reference range.

Further information for GPs

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

NICE (2019) Prostate cancer: diagnosis and management. 🖱 https://www.nice.org.uk/guidance/ng131

Information for patients

Cancer Research UK ☎ 0808 800 4040 🖱 www.cancerresearchuk.org
Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk
NHS Choices 🖱 www.nhs.uk/conditions/prostate-cancer/psa-testing/
Prostate Cancer UK ☎ 0800 074 8383 🖱 https://prostatecanceruk.org/
The National Federation of Prostate Cancer Support Groups ☎ 0800 035 5302 🖱 http://tackleprostate.org

Treatment of prostate cancer

Symptomless local disease Treatment is controversial. There are 2 arguments:

Benefits of treatment are outweighed by risks	*Or*	Aggressive treatment before spread is the only way to ensure cure.

>50% of ♂ >50y who die from other causes are found postmortem to have prostate cancer—prostate cancer kills only a small minority of men who have it. The personal and economic cost of treating men whose cancer would never have caused them any problems must be considered.

Options

- **Watchful waiting or active surveillance** Monitor with PSA and regular rectal examination. ↑ in PSA or size of nodule triggers active treatment. A large UK trial published in 2016 has found active monitoring is as effective as surgery and radiotherapy, in terms of survival at 10 years with less side effects. Progression rates are higher in patients with poorly differentiated cancer. Some men find the uncertainty of waiting difficult to cope with
- **Radical prostatectomy** Has potential for cure, but in the age group most affected by prostate cancer, mortality is 1.4%. Other common complications: impotence (50%), incontinence (25%)
- **Radiotherapy** May not be effective—persistent cancer is found in 30% on biopsy. Brachytherapy (radioactive treatment in implanted seeds or wires) has proven efficacy in early prostate cancer
- **Hormone treatment** No convincing evidence that this gives survival benefit in early disease
- **Others** Minimally invasive treatments, e.g. cryo- or microwave therapy

Symptomatic disease 30% 5y survival. Hormone manipulation is the mainstay of treatment and gives 80% ↓ in bone pain, PSA, or both and a lower incidence of serious complications (e.g. spinal cord compression) if treatment starts at the time of diagnosis. *Options*:

Luteinizing hormone-releasing hormone (LHRH) analogues e.g. goserelin—sc injection every 4–12wk (depending on the preparation used). Testosterone levels ↓ to levels of castrated men in <2mo. *Side effects*: impotence, hot flushes, gynaecomastia, local bruising and infection around injection site. When starting LHRH analogues, LH level initially ↑ which can cause increased tumour activity or 'flare'. Counteracted by prescription of anti-androgens (e.g. flutamide) for a few days before administration of the first dose of LHRH and concurrently for 3wk. Response in most patients lasts for 12–18mo.

Anti-androgens e.g. cyproterone acetate, flutamide, bicalutamide. Do not suppress androgen production completely. Used to prevent side effects due to testosterone flare during initiation of LHRH analogues, as monotherapy (e.g. bicalutamide 150mg od), and in combination with LHRH analogues to produce maximum androgen blockade.

Surgical castration ↓ testosterone secretion permanently without the need for medication. However, rarely used.

Bony metastases In addition to hormone therapy, local radiotherapy and corticosteroids are used for bone pain. Radioactive strontium ↓ the number of new sites of bone pain developed. Mean survival <5y.

Hormone-resistant disease No agreed treatment. Involve the multidisciplinary team including urology, oncology, and palliative care. Dexamethasone 0.5mg daily or docetaxel may be helpful.

Factors affecting prognosis of prostate cancer

Stage

Tumour		Lymph nodes?		Metastases?	
T1	Impalpable	N0	No	M0	No spread outside the pelvis
T2	Tumour completely within the prostate gland	N1	**1** +ve LN <2cm diameter	M1	Spread outside the pelvis
T3	Tumour has breached the capsule of the prostate	N2	>1 +ve LN or 1 LN of 2–5cm diameter		
T4	Spread within the pelvis e.g. to bladder or bowel	N3	Any +ve LN >5cm diameter		

Gleason score Histological grade. Cells are graded 1–5 the less differentiated they are. The 2 areas of the biopsy with the highest grade cells are added together. Low-grade tumours likely to grow slowly have low scores (2–4); high-grade tumours have high scores (7–10).

Age Older patients with low-grade tumours are likely to die from something other than their prostate cancer.

PSA
- PSA >40: high chance of nodal or metastatic spread
- PSA >100: metastatic spread is very likely

Prognosis 5y survival rates for tumour stage:
- 1 or 2—tumour confined within the prostate (65–98%)
- 3—tumour has breached the capsule of the prostate (60%)
- 4—spread to LNs, within the pelvis, or elsewhere (20–30%)

Further information

Hamdy FC, et al. (2016). 10-Year outcomes after monitoring, surgery, or radiotherapy for localized prostate cancer. *NEJM* **371**:415–24.

NICE (2019) Prostate cancer: diagnosis and management. https://www.nice.org.uk/guidance/ng131

Conditions of the penis

Posterior urethral valves Folds of mucosa inhibit or block passage of urine causing urethral, bladder, ureter, and renal pelvis dilatation.

Presentation Usually detected on antenatal USS. Can present in neonates with urinary retention or dribbling urine + distended bladder, UTI or uraemia, or later in childhood with recurrent UTI or incontinence.

Investigation and management MCUG confirms diagnosis. In all cases refer to urology for surgical disruption of the valves.

Hypospadias 1 in 400 male births. The urethral meatus opens on the ventral side of the penis. There is often hooding of the foreskin and ventral flexion of the penis. Refer to urology. Treated with corrective surgery, ideally preschool.

Non-retractile foreskin Usually noted by parents. May be history of recurrent balanitis. *Examination*: foreskin adherent.

Management Age <4y—do nothing unless recurrent balanitis. If >4y and/or recurrent balanitis, consider treatment with topical steroids (e.g. betamethasone 0.1% od) for 3–4mo. If ineffective, refer to paediatric surgery for circumcision.

Phimosis Foreskin obstructs urine flow. Common in small children. Time usually obviates the need for circumcision. Treat as for non-retractile foreskin if recurrent balanitis.

Peyronie's disease Hard lumps in the shaft of the penis. Unknown cause. 4% ♂ >40y. 1 in 3 have pain/bending of the penis when erect. Associated with erectile dysfunction (➔ p. 754). 5% have Dupuytren's contracture.

Management Reassurance usually suffices. No proven medical treatments. Refer to urology for surgery if pain or severe bending on erection so that intercourse is not possible.

Paraphimosis Foreskin is retracted then (because of oedema) unable to be replaced. Commonly occurs in catheterized patients when the catheter is changed.

Management Try to replace foreskin using ice packs (↓ swelling) and lubrication (e.g. K-Y® Jelly). If unable to replace the foreskin, admit for surgery.

Balanitis Acute inflammation of glans and foreskin. Common organisms—staphylococci, streptococci, coliforms, *Candida*. Can occur at any age. Most common in young boys when associated with non-retractile foreskin/phimosis. In elderly patients consider DM.

Management Oral antibiotics (e.g. flucloxacillin) or topical antifungals (e.g. clotrimazole). If recurrent or secondary to phimosis consider referral for circumcision.

Balanitis xerotica et obliterans Chronic fibrosing condition of the foreskin which may become adherent to the glans. Treatment is with topical steroid creams, e.g. betamethasone 0.1%. Consider referral for circumcision.

Trauma to the foreskin Torn frenulum—seen after poorly lubricated intercourse or if caught in a zip. No treatment required. If recurrent, consider referral for circumcision.

Erectile dysfunction ➔ p. 754

Priapism Persistent painful erection not related to sexual desire.

Cause Medication for erectile dysfunction, idiopathic, leukaemia, sickle cell disease, or pelvic tumour.

Management Ask the patient to climb stairs (arterial 'steal' phenomenon), apply ice packs. If unsuccessful, refer to A&E for aspiration of corpora. Rarely surgery is needed.

Erythroplasia of Queyrat Premalignant condition of glans. Moist velvety-looking patches. Refer to urology. Treatment is surgical.

Carcinoma of the penis Squamous cell carcinoma (95%) or malignant melanoma. Usually elderly men. Rare in the UK.

Management[N] Refer urgently patients with symptoms or signs of penile cancer. These include:

- Progressive ulceration in the glans, prepuce, or skin of the penile shaft
- Mass in the glans, prepuce, or skin of the penile shaft

❗ Lumps within the corpora cavernosa can indicate Peyronie's disease, which does not require urgent referral.

Penile discharge Associated with urethritis, e.g. due to chlamydia or gonorrhoea. Refer to GUM clinic.

Further information

BASHH (2008) Management of balanoposthitis. 🖱 https://www.bashhguidelines.org/media/1077/2062.pdf

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Testicular disease

Testicular pain Treat the cause:

- Epididymo-orchitis
- Torsion of the testis
- Trauma and haematoma formation
- Varicocele
- Testicular tumour (rarely painful)

Torsion of the testis Peak age 15–30y. Presents with sudden-onset, severe scrotal pain. May be associated with right iliac fossa pain, nausea, and vomiting. *Examination*: tender, hard testis riding higher than contralateral testis. Admit urgently to surgical/urology team

Torsion of the hydatid of Morgagni Small embryological remnant at the upper pole of the testis. Presents similarly to torsion of the testis. Refer as an emergency to exclude torsion of the testis.

Epididymo-orchitis Inflammation of the testis and epididymis due to infection. May occur at any age. The most common viral cause is mumps. The most common bacterial causes are *Chlamydia* or gonococci (<35y) and coliforms (>35y). Chronic infection with TB or syphilis is rare.

Presentation Acute-onset pain in testis; swelling and tenderness of testis/epididymis; fever ± rigors; may be urethritis, dysuria and/or ↑ frequency.

Management May be difficult to distinguish from torsion of the testis. If in doubt, admit for urology/surgical opinion. Otherwise investigate and treat for the underlying cause.

Testicular lumps and swellings Figure 13.3

Hydrocele Collection of fluid in tunica vaginalis. Occurs at any age.

- 1° hydrocele—no predisposing cause in scrotum
- 2° hydrocele—reaction to pathology in testis or covering (infection, tumour, torsion). In adults presenting with hydrocele, always consider impalpable tumour beneath

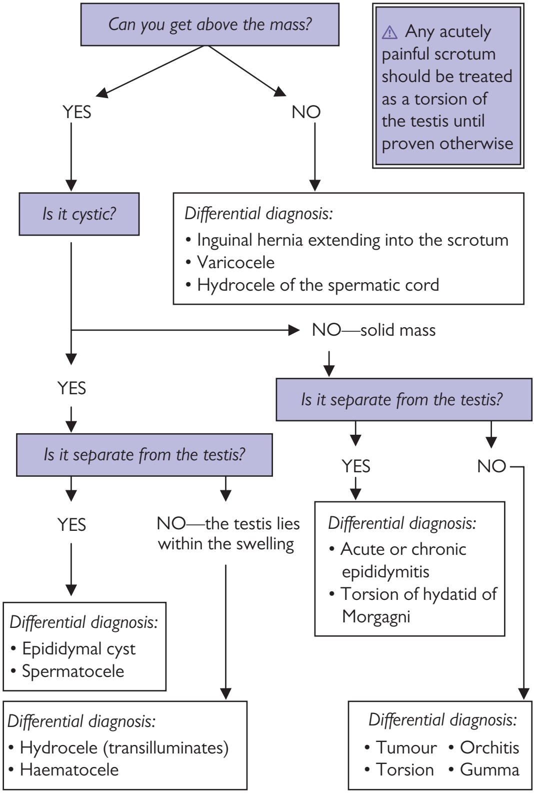
Presentation Swelling in the scrotum. The examiner should be able to get above swelling. Smooth surface, transilluminates, testis is within the swelling and not palpable separately.

Management Investigation is not required in children; refer adults for USS if testis is not palpable. Options for adults:

- Conservative management—reassurance—small hydroceles
- Tapping—may be suitable for large hydroceles where surgery is inappropriate—2° infection and recurrence are common
- Surgery—refer to urologist

Hydroceles in children are usually congenital. May be unilateral or bilateral. Most resolve spontaneously in the first year of life. Refer to urology if persists >1y.

Hydrocele of the cord Arises in part of the processus vaginalis in the spermatic cord above the testis. Rounded lump which slips up and down the inguinal canal. No action needed.


Can you get above the mass?
YES
NO
⚠ Any acutely painful scrotum should be treated as a torsion of the testis until proven otherwise
Is it cystic?
Differential diagnosis:
• Inguinal hernia extending into the scrotum
• Varicocele
• Hydrocele of the spermatic cord
NO—solid mass
YES
Is it separate from the testis?
Is it separate from the testis?
YES
NO
YES
NO—the testis lies within the swelling
Differential diagnosis:
• Acute or chronic epididymitis
• Torsion of hydatid of Morgagni
Differential diagnosis:
• Epididymal cyst
• Spermatocele
Differential diagnosis:
• Hydrocele (transilluminates)
• Haematocele
Differential diagnosis:
• Tumour • Orchitis
• Torsion • Gumma

Referral guidelines[N]

- Refer urgently patients with a swelling or mass in the body of the testis
- Consider an urgent USS in men with a scrotal mass that does not transilluminate and/or when the body of the testis cannot be distinguished

Figure 13.3 Diagnosis of testicular lumps

Further information

BASHH (2010) Management of epididymo-orchitis. https://www.bashhguidelines.org/media/1062/3546.pdf

NICE (2015, updated 2017) Suspected cancer: recognition and referral. www.nice.org.uk/guidance/ng12

Haematocele Damage to the testis (e.g. due to a direct blow, vasectomy), can result in the testis rupturing and the tunica vaginalis filling with blood. Refer as an emergency for urological assessment.

Varicocele Collection of varicose veins in the pampiniform plexus of the cord and scrotum. Can be 2° to obstruction of the testicular veins in the abdomen. L > R. Associated with infertility (thought due to ↑ temperature of testis). Presents with a dull ache in the testis especially at the end of the day or after exercise. Usually visible when the patient is standing. No treatment is needed—reassure. Occasionally surgery or radiological embolization may help if symptoms are severe.

Epididymal cyst Common and often multiple. Found in middle-aged/elderly men. Usually presents when the patient finds a painless lump.

- Examination Smooth-walled cysts in epididymis (palpable above and behind testis), often bilateral
- Investigation If unsure of diagnosis refer for USS
- Management Reassurance. Refer to urology if painful

Spermatocele Cyst containing sperm. Typically situated in the head of the epididymis—more rarely in the spermatic cord. Clinically presents in the same way as epididymal cyst. Management is the same.

Testicular gumma ➔ p. 725

Benign testicular tumours Rare (<2% tumours). Sertoli cell adenomas; Leydig cell adenomas. Produce sex hormones and cause feminization/masculinization respectively. Refer.

Testicular cancer Most common malignancy in men age 20–34y. Devastating disease as sufferers tend to be young and fit and do not expect to be ill. Screening is not effective. Education to ensure men check their testes for lumps regularly and present early is preferable.

Risk factors Undescended testes—bilateral undescended testis → 10× ↑ risk; past history of testicular cancer—4% risk 2nd cancer.

Presentation Painless lump in testis; occasionally testicular pain or hydrocele; may present with metastases—back pain/dyspnoea.

Management Testicular lumps are tumours until proven otherwise. Refer for urgent urological opinion. USS can help diagnosis but *do not* delay referral. Definitive diagnosis is only made at biopsy. Specialist treatment depends on tumour type and extent (Table 13.10). Sperm banking is routinely offered in case of ↓ fertility due to treatment.

❗ Children conceived by men treated for testicular cancer are not at ↑ risk of congenital abnormality.

Empty scrotum If the scrotum has never contained a testis, it is hypoplastic. If the scrotum has contained a testis in the past, it is normally developed but empty.

Causes of an empty scrotum Undescended or retractile testis; surgical removal, e.g. for torsion, trauma, or tumour; testicular atrophy (e.g. due to mumps or trauma); ambiguous genitalia; testicular agenesis—diagnosis of exclusion.

Table 13.10 Types and features of testicular cancer

	Seminoma (60%)	Teratoma
Typical age	30–40y	<30y
Tumour markers	None	β-HCG AFP LDH—correlates with volume of metastatic disease
Nature of tumour	Solid	Solid/cystic components. 40% occur within seminomas. Mixed tumours are treated like teratomas
Growth speed	Slow growing	Fast growing—can ×2 in size in days
Stage of presentation	90% stage 1 (tumour confined to testis)	60% stage 1 (tumour confined to testis)
Treatment	Treated with inguinal orchidectomy + radiotherapy Relapses are treated with chemotherapy. More advanced disease is treated with radio- or chemotherapy	Treatment of stage 1 disease is with inguinal orchidectomy and surveillance of tumour markers. 25% relapse in <18mo Treatment of relapses and metastatic disease is with chemotherapy
Survival	98% 5y survival for stage 1 disease. Overall >85% 5y survival	Prognosis depends on stage and degree of differentiation

Carcinoma of the scrotal skin SCC or melanoma. Uncommon <50y. Painless lump/ulcer of the scrotal skin ± enlarged inguinal LNs. If suspected, refer urgently to urology or dermatology.

Fournier's gangrene Necrotizing fasciitis of the scrotal skin and/or penis. Patients are usually elderly and often have a hydrocoele. Starts as a black spot and spreads rapidly. Early diagnosis is critical to survival so, if suspected admit as an acute urological emergency. Treatment is with surgical debridement and IV antibiotics.

Undescended testis Affects 2–3% of ♂ neonates—but most descend during the 1st year. Refer those that do not for surgical descent/fixation to avoid ↑ risk of malignancy and later infertility.

Retractile testis Usually young boys with active cremasteric reflex. No treatment needed.

Examination Scrotum is usually well developed. Try to find the testis and milk it down into scrotum. May be found anywhere from the scrotum to the internal inguinal ring. If not found or you are unable to bring the testis down into the scrotum assume it is undescended.

Information and support for patients with testicular cancer

Cancer Research UK ☎ 0808 800 4040 🖱 www.cancerresearchuk.org
Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk

Chapter 14

Musculoskeletal problems

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Symptoms of musculoskeletal disease

Bone pain *Consider*:
- Fracture—due to injury, stress fracture, or pathological fracture
- Arthritis—referred pain from affected joints
- Malignancy—primary bone malignancy, haematological malignancy, e.g. multiple myeloma, or secondaries (usually from breast, prostate, lung, thyroid, kidney—more rarely bowel, melanoma)
- Benign bone tumour
- Infection—osteomyelitis or joint infection
- Metabolic causes e.g. hypercalcaemia

Pain in one joint Common. Ask:

Is the problem articular or periarticular?
- Articular disease (e.g. osteoarthritis) is suggested by joint line tenderness and pain at the end of the range of movement in any direction
- Periarticular problems (e.g. ligamentous injury)—point tenderness over the involved structure, and pain exacerbated by movements

If articular Is the problem inflammatory or mechanical? Look for:
- Signs of inflammation—warmth, redness, effusions—may indicate joint infection or inflammatory arthritis
- Features of a mechanical problem—locking/catching, e.g. cartilage tear

If periarticular Which structure is causing pain? *Options*: bursa; tendon; tendon sheath; ligament; soft tissue; bony epiphysis/metaphysis

⚠ *Red flags* Features which should prompt early/urgent referral:
- Inflamed joint with associated fever or constitutional disturbance—beware of septic arthritis
- Any joint which is 'locked' or so painful that movement is impossible
- Severe pain at rest or at night
- Pain that gets relentlessly worse over a period of days or weeks

Pain in multiple joints Differentiate between articular or periarticular disease, and whether the condition is inflammatory or not. Screening with blood tests (ESR or CRP, FBC, ± autoimmune profile) may help. Look for the pattern of disease, e.g. joint sites involved; other symptoms/signs.

Common arthropathies ➔ pp. 486–99
- Osteoarthritis
- Rheumatoid arthritis
- Ankylosing spondylitis
- SLE
- Reactive arthritis
- Psoriatic arthritis
- Enteropathic arthropathy
- Gout or pseudogout
- Sjögren's syndrome
- Malignancy

⚠ *Red flags* Features which should prompt early/urgent referral:
- Severe systemic symptoms—high fevers, significant weight loss, or a very ill patient (suggests rheumatoid arthritis, sepsis, or malignancy)
- Focal systemic signs, e.g. rashes, nodules, or GI disturbances
- Severe pain and/or inability to function

Back pain ➲ p. 450 **Neck pain** ➲ p. 448

Joint stiffness Clarify what the patient means—stiffness may refer to either a loss of range of movement ± pain and/or pain on movement (e.g. following unaccustomed exercise). *Morning stiffness* is pain that eases on movement and is characteristic of inflammatory arthritis, particularly RA.

Joint swelling Common. Ask: what does the swelling feel like?

- Hard/bony swelling or deformity—usually osteoarthritis (e.g. osteophytes) but may result from destructive arthropathy (e.g. inflammatory arthritis, Charcot's joint), injury, or heterotopic calcification
- Soft and fluctuant—suggests joint effusion as a result of inflammation or infection within the joint
- Soft and non-fluctuant—soft tissue swelling resulting from injury, infection, or inflammation
- Firm and non-fluctuant—suggests synovial thickening resulting from inflammatory arthritis

Deformity Abnormal shape. May be temporary (e.g. soft tissue swelling after injury, new fracture) or permanent (e.g. congenital malformation, Paget's disease, osteoarthritis). Look for the underlying cause.

Dystonia ➲ p. 522 **Chest deformity** ➲ p. 268
Short stature ➲ p. 870 **Excess height** ➲ p. 871

Myalgia Isolated myalgia can be a result of overuse or soft tissue injury. Generalized myalgia is associated with many diseases including:

- Infection
- Fibromyalgia
- Chronic fatigue syndrome
- Myositis associated with statin
- PAN
- Granulomatosis with polyangiitis

Children with musculoskeletal pain of unknown cause

Take a history and examine carefully to exclude other causes. Investigate further with FBC, blood film, ESR, ± X-ray if bone pain, rest pain, or persistent or unexplained back pain[N]. If no cause is found, treatment is with analgesia and reassurance. Advise to return for reassessment ± orthopaedic referral if pain worsens, continues >6wk, changes in nature, or other symptoms develop.

Nocturnal musculoskeletal pains (growing pains) Episodic, muscular pains, usually in the legs, lasting ~30min and waking the child from sleep. Rubbing the limb brings rapid relief. There is no pain or disability in the morning. Diagnosis can be made on history if there are no associated symptoms and examination is normal. If in doubt, check FBC and ESR—which should be normal. In most cases reassurance ± analgesia are all that is needed. In resistant cases, physiotherapy may help.

The limping child ➲ p. 465

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Neck pain

⚠ **Neck trauma** Any significant cervical trauma requires neck immobilization with a hard collar and referral to A&E for cervical spine X-rays to exclude vertebral fracture/instability that could threaten the spinal cord.

Neck pain is common (lifetime incidence 50%) and contributes to 2% of GP consultations. Prevalence is highest in middle age. Most neck pain is acute and self-limiting (within days/weeks) but 1 in 3 have symptoms lasting >6mo or recurring pain.

History

- Pain Onset, site, radiation, aggravating and relieving factors, timing
- Stiffness Timing—continuous? worse in the mornings?
- Deformity e.g. torticollis. Onset, changes
- Neurological symptoms Numbness, paraesthesiae, weakness
- Other symptoms Weight ↓, sweats, bowel/bladder dysfunction (consider spinal cord compression)

❗ Pain is often poorly localized and neck problems commonly present with shoulder pain and/or headache (cervicogenic headache).

Examination

- Look Posture; deformity, e.g. torticollis, asymmetry of scapulae; arms and hands—wasting, fasciculation? leg weakness?
- Feel Tenderness? Midline tenderness may be due to supraspinous or spinous process damage following a whiplash injury. Paraspinal tenderness ± spasm radiating into the trapezius ± crepitation is common with cervical spondylosis
- Move/measure Normal ranges: flexion/extension—130° total range; lateral flexion—45° in each direction from a neutral position; rotation—80° in each direction from a neutral position
- Neurology Weakness in the upper limbs in a segmental distribution, with loss of dermatomal sensation and altered reflexes indicates a root lesion (Table 14.1). If cervical cord compression is suspected, examine the lower limbs looking for upgoing planters and hyperreflexia

Cervical spondylosis Degenerative disease of the cervical spine. May cause pain but minor changes are normal (especially >40y) and usually asymptomatic. Pain is generally intermittent and related to activity. Examination reveals ↓ neck mobility. Severe degeneration can cause nerve root signs. Treat with analgesia. X-ray only if conservative measures fail, troublesome pain, nerve root signs, or psoriasis (?psoriatic arthropathy).

Nerve root irritation or entrapment 2° to degeneration, vertebral displacement/collapse, disc prolapse, local tumour, or abscess. Causes neck stiffness, pain in arms or fingers, ↓ reflexes, sensory loss, and ↓ power. Spurling's test (neck extension + rotation to the affected side followed by downwards, axial pressure) reproduces symptoms. Determine level of entrapment clinically (Table 14.1). Treat with analgesia. X-ray cervical spine—lateral/views. Refer for physiotherapy ± further investigations (e.g. MRI) if conservative management fails and there is objective evidence of a root lesion

Table 14.1 Neurology associated with cervical nerve root entrapment

Root	Sensory changes	Motor weakness	Reflex changes
C5	Lateral arm	Shoulder abduction/flexion Elbow flexion	Biceps
C6	Lateral forearm Thumb Index finger	Elbow flexion Wrist extension	Biceps Supinator
C7	Middle finger	Elbow extension Wrist flexion Finger extension	Triceps
C8	Medial side of lower forearm Ring and little fingers	Finger flexion	None
T1	Medial side of upper forearm	Finger abduction/adduction	None

⚠ **Red flags** Refer as an emergency if signs of spinal cord compression:

- Root pain and lower motor neurone signs at the level of the lesion *and*
- Spastic weakness, brisk reflexes, upgoing plantars, loss of coordination and sensation below the lesion

Spasmodic torticollis (wry neck) Common. Sudden-onset, painful stiff neck due to spasm of trapezius and sternocleidomastoid muscles. Self-limiting. Heat, gentle mobilization, muscle relaxants, and analgesia can speed recovery. Often caused by poor posture—e.g. computer-seating position; carrying heavy uneven loads.

Thoracic outlet syndrome (TOS) Caused by neurovascular compression as nerves/blood vessels pass through the thoracic outlet (ring formed by the scalene muscles, first rib, and clavicle). May present with shoulder/arm pain, weakness/paraesthesia ± thenar/hypothenar wasting. Radial pulse may be weak. Refer to upper limb orthopaedic surgeon for assessment if suspected.

Cervical rib Congenital C7 costal process enlargement. Usually asymptomatic but can cause TOS. X-ray may show cervical rib—but symptoms are sometimes due to fibrous bands not seen on X-ray.

Whiplash injuries Neck pain caused by stretching/tearing of cervical muscles and ligaments due to sudden extension of the neck—often due to a RTA. Pain and ↓ neck mobility typically starts several hours or days after injury. Pain may radiate to shoulders, arms, and head.

Management Examine to exclude bony tenderness requiring X-ray. Treat with analgesia and early mobilization. A soft collar may help initially but avoid long-term use. Recovery is often slow; 40% suffer long-lasting symptoms. As a general rule, the quicker symptoms develop, the longer they take to disappear. Early physiotherapy, if available, ↑ recovery rate. Psychological problems and medico-legal issues may affect progress.

Low back pain

Definitions

- **Acute low back pain** New episode of low back pain of <6wk duration. Common—lifetime prevalence 58%
- **Chronic low back pain** Back pain lasting >3mo

Causes of back pain

Table 14.2

History

Ask about:

- **Circumstances of pain**—history of injury; duration
- **Nature/severity of pain**—pain/stiffness mainly at rest/at night, easing with movement suggests inflammation (e.g. discitis, spondyloarthropathy)
- **Associated symptoms**—numbness, weakness, bowel/bladder symptoms
- **PMH**—past illnesses (e.g. cancer), previous back problems
- **Exclude pain not coming from the back** (e.g. GI/GU pain; AAA)

Examination

- Deformity, e.g. kyphosis (typical of ankylosing spondylitis), loss of lumbar lordosis (common in acute mechanical back pain), scoliosis
- Palpate for tenderness, step deformity, and muscle spasm
- Assess flexion, extension, lateral flexion, and rotation while standing
- Ask to lie down—this gives a good indication of severity of symptoms
- In lower limbs look for muscle wasting and check power, sensory loss, and reflexes (knee jerk and ankle jerk)—Table 14.3. Assess straight leg raise (SLR)—sciatica is present if SLR on one side elicits back/buttock pain (usually ipsilateral) compared to SLR on the other side

⚠ Red flags

- <20 or >55y
- Non-mechanical pain
- Pain that worsens when supine
- Night-time pain
- Thoracic pain
- Past history of cancer
- AAA
- HIV
- Immune suppression
- IV drug use
- Taking steroids
- Unwell
- Weight ↓
- Widespread neurology
- Structural deformity

Management of acute back pain in the community Triage according to history and examination—Figure 14.1, ➔ p. 452

For patients who do not require immediate referral Consider analgesia, e.g. NSAIDs (for short time + consider gastroprotection); weak opioids (± paracetamol). Use Keele STarT Back screening tool (Box 14.1):

- If total score ≤3, explain likely natural history of the pain and advise to avoid bed rest and maintain normal activities as far as possible (↓ chance of chronic pain). Suggest self-help exercises
- If total score is ≥4, check question 5–9 sub-score:
  - If ≤3—if not resolved in 4wk, refer for physical therapy. Options include: back exercise classes, physiotherapy, chiropractic, osteopathy or acupuncture, if available
  - If ≥4—if not resolved in 4wk, refer directly for specialist intervention—sooner if worsening or severe pain
- In all cases, challenge any '*yellow flag*' factors (Figure 14.1, ➔ p. 452) that may inhibit recovery and delay return to normal functioning

Table 14.2 Causes of back pain: age suggests the most likely cause

Age (y)	Causes		
15–30	• Postural • Mechanical • Prolapsed disc	• Trauma • Fracture • Ankylosing spondylosis	• Spondylolisthesis • Pregnancy
30–50	• Postural • Prolapsed disc	• Spondyloarthropathies • Discitis	• Degenerative joint disease
>50	• Postural • Degenerative • Paget's disease	• Malignancy (lung, breast, prostate, thyroid, kidney) • Myeloma	• Osteoporotic collapse
Other causes	• Spinal stenosis • Cauda equina tumour	• Abdominal aortic aneurysm	• Spinal infection • Referred pain

Table 14.3 Neurology with lumbosacral nerve root entrapment

Root	Sensory changes	Motor weakness	Reflex changes
L2	Front of thigh	Hip flexion/adduction	None
L3	Inner thigh	Knee extension	Knee
L4	Inner shin	Knee extension Foot dorsiflexion	Knee
L5	Outer shin Dorsum of foot	Knee flexion Foot inversion Big toe dorsiflexion	None
S1	Lateral side of foot/sole	Knee flexion Foot plantarflexion	Ankle

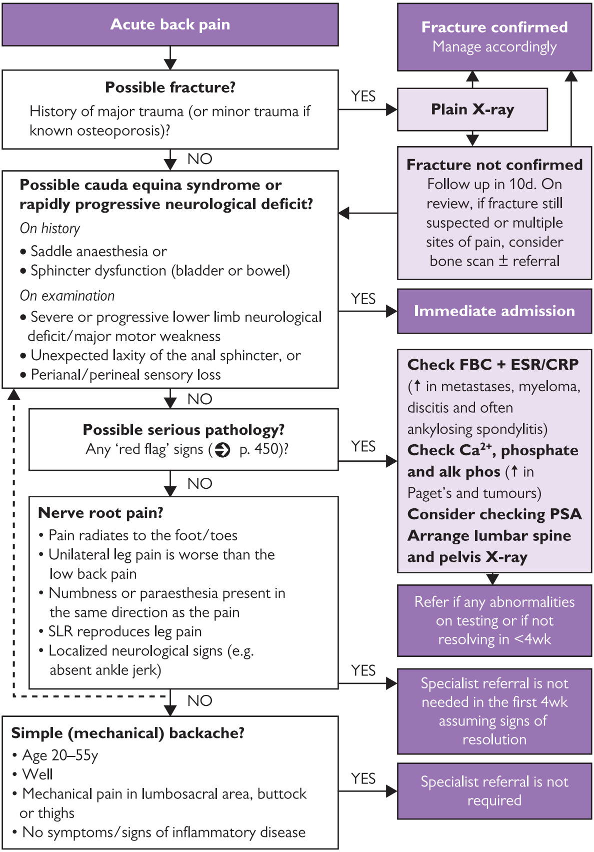
Box 14.1 Keele STarT Back Pain Scoring Tool

Ask patients to consider the following statements and state whether they agree or disagree with them. *Thinking about the past 2wk*:

1. My back pain has spread down my leg(s) at some time in the last 2wk
2. I have had pain in the shoulder or neck at some time in the last 2wk
3. I have only walked short distances because of my back pain
4. In the last 2wk, I have dressed more slowly than usual because of back pain
5. It's not really safe for a person with a condition like mine to be physically active
6. Worrying thoughts have been going through my mind a lot of the time
7. I feel that my back pain is terrible and it's never going to get any better
8. In general I have not enjoyed all the things I used to enjoy

If the patient agrees with a statement, score 1; if disagrees, score 0.

9. Overall, how bothersome has your back pain been in the last 2wk?
    - *Not at all, slightly or moderately—score 0*
    - *Very much or extremely—score 1*


Acute back pain
Possible fracture?
History of major trauma (or minor trauma if known osteoporosis)?
YES
Plain X-ray
Fracture confirmed
Manage accordingly
Fracture not confirmed
Follow up in 10d. On review, if fracture still suspected or multiple sites of pain, consider bone scan ± referral
NO
Possible cauda equina syndrome or rapidly progressive neurological deficit?
On history
• Saddle anaesthesia or
• Sphincter dysfunction (bladder or bowel)
On examination
• Severe or progressive lower limb neurological deficit/major motor weakness
• Unexpected laxity of the anal sphincter, or
• Perianal/perineal sensory loss
YES
Immediate admission
NO
Possible serious pathology?
Any 'red flag' signs (➜ p. 450)?
YES
Check FBC + ESR/CRP
(↑ in metastases, myeloma, discitis and often ankylosing spondylitis)
Check Ca2+, phosphate and alk phos (↑ in Paget's and tumours)
Consider checking PSA
Arrange lumbar spine and pelvis X-ray
Refer if any abnormalities on testing or if not resolving in <4wk
NO
Nerve root pain?
• Pain radiates to the foot/toes
• Unilateral leg pain is worse than the low back pain
• Numbness or paraesthesia present in the same direction as the pain
• SLR reproduces leg pain
• Localized neurological signs (e.g. absent ankle jerk)
YES
Specialist referral is not needed in the first 4wk assuming signs of resolution
NO
Simple (mechanical) backache?
• Age 20–55y
• Well
• Mechanical pain in lumbosacral area, buttock or thighs
• No symptoms/signs of inflammatory disease
YES
Specialist referral is not required

Yellow flags—psychosocial barriers to back pain recovery

- Belief that pain and activity are harmful
- Sickness behaviours, e.g. extended rest
- Social withdrawal
- Emotional problems, e.g., low mood, depression, anxiety, or stress
- Problems and/or dissatisfaction at work
- Problems with claims/compensation/time off work
- Overprotective family; lack of support
- Inappropriate expectations of treatment

Figure 14.1 Triage of acute back pain

❗ *Do not X-ray routinely* X-rays require a high radiation dose and clinically meaningful findings are rare. *Exceptions*:

- Young (<25y)—X-ray sacroiliac joints to exclude ankylosing spondylitis
- Elderly—if vertebral collapse/malignancy suspected
- History of trauma

⚠ **Cauda equina syndrome** Compression of the cauda equina below L2, e.g. by disc protrusion at L4/5. Presents with:

- Numbness of the buttocks and backs of thighs
- Urinary/faecal incontinence
- Lower motor neurone weakness:
  - L4—loss of dorsiflexion of the foot (and toes—L4/5)
  - S1—loss of ankle reflex, plantarflexion and eversion of the foot

Management Refer/admit as a neurological emergency. Rapid surgical intervention ↑ the chance of full motor and sphincter recovery.

⚠ **Spinal cord compression** Affects 5% of cancer patients—70% in the thoracic region. Maintain a *high* level of suspicion if history of cancer and new back pain—especially if known bony metastases or tumour likely to metastasize to bone. Presents with:

- Back pain, worse on movement—often appears before neurology
- Neurological symptoms/signs—can be non-specific, e.g. constipation, weak legs, urinary hesitancy. Lesions above L1 (lower end of spinal cord) produce upper motor neurone signs (e.g. ↑ tone/reflexes) and a sensory level; lesions below L1 produce lower motor neurone signs (↓ tone/reflexes) and peri-anal numbness (cauda equina syndrome)

Management Prompt treatment (<24–48h from first neurological symptoms) is needed; once paralysed, <5% walk again. Treat with oral dexamethasone 16mg/d and refer for same day assessment and surgery/radiotherapy unless in final stages of disease.

Osteoporotic vertebral collapse ➔ p. 482

Scoliosis Lateral curvature of the spine associated with rotation of vertebrae ± ribs or wedging of vertebrae. Early treatment prevents progression and complications, e.g. cardiopulmonary disturbance. *Causes*:

- Idiopathic
- Congenital (butterfly vertebra)
- Infection—TB of spine
- Metabolic, e.g. bone dysplasias
- Neuromuscular problems, e.g. cerebral palsy, neurofibromatosis, Friedreich's ataxia, muscular dystrophy, polio
- Trauma → damage in vertebral growth plate and uneven growth
- Neoplasm 1°, 2°, or as a result of radiotherapy

Clinical features Difference in shoulder height; spinal curvature; difference in the space between the trunk and upper limbs. ❗ Scoliosis which disappears on bending is postural and of no clinical significance.

Management In all cases where structural scoliosis is suspected, refer to orthopaedics—urgently if associated with pain, especially at night.

Further information

NICE (2016) Low back pain and sciatica in over 16s. 🖱 www.nice.org.uk/guidance/ng59

Patient information and support

Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org

Shoulder problems

History

- **Pain and stiffness** Joint pain is felt anteriorly and may radiate down the arm; pain on top of the shoulder suggests acromioclavicular joint problems or cervical spine disorders. ❗ Pain in the shoulder may be referred from the neck, heart, mediastinum, or diaphragm
- **Deformity** Swelling of the shoulder; prominence of the acromioclavicular joint; winging of the scapula
- **Loss of function** Difficulty reaching behind back (e.g. doing up bra strap), brushing hair, or dressing

Examination

- **Look** Posture; asymmetry; muscle wasting; swelling (large effusions can be seen anteriorly); scars
- **Feel** Tenderness; warmth; swelling; crepitus
- **Move/measure** Compare sides. Check range of movement; complex movements (e.g. hands behind head, arm across front of chest to top of opposite shoulder, hand behind back); power

General rules Intra-articular disease—painful limitation of movement in all directions; tendonitis—painful limitation of movement in one plane only; tendon rupture or neurological lesions—painless weakness.

⚠ *Red flags*

- Past history of carcinoma
- Constitutional symptoms, e.g. fever, chills, or unexplained weight ↓
- Recent bacterial infection
- IV drug use
- Immune suppression
- Constant/worsening rest pain
- Structural deformity

Causes of a stiff, painful shoulder joint

- Adhesive capsulitis—1° or 2° to DM or intrathoracic pathology
- Inflammation—inflammatory arthritis (e.g. RA, psoriatic), infection
- Osteoarthritis
- Prolonged immobilization, e.g. hemiplegia, strapping after dislocation
- Polymyalgia rheumatica

Shoulder osteoarthritis Often occurs after trauma. Less common than knee or hip OA. May be associated with crystal-induced inflammation and 2° causes of OA (e.g. gout, haemochromatosis). Imaging for synovitis (USS/MRI) is important to exclude disease that may benefit from steroid injection. Consider referral for shoulder replacement if severe.

Frozen shoulder (adhesive capsulitis) Overdiagnosed in primary care. Affects patients aged 40–60y. Painful, stiff shoulder with global limitation of movement—notably external rotation. Pain is often worse at night. Cause unknown but ↑ in people with DM, and those with intra-thoracic pathology (MI, lung disease) or neck disease.

Management If not known to have diabetes, check HbA1c. NSAIDs, physiotherapy, and local steroid injection can all be helpful. May take >1y to recover and long-term outcome is uncertain. If restricted movements are slow to return, consider orthopaedic referral.

Rotator cuff injury The shoulder is the most mobile joint in the body and relies on the musculotendinous rotator cuff to maintain stability. Disorders of the rotator cuff account for most shoulder pain.

- **Acute tendinitis** Often due to excessive use/trauma in patients <40y. Presents with severe pain in the upper arm. Patients hold the arm immobile and are unable to lie on the affected side. Usually starts to resolve spontaneously after a few days. In middle age can be caused by inflammation around calcific deposits—requires steroid injection
- **Rotator cuff tears** May accompany subacromial impingement; difficult to diagnose clinically unless large—suspect if recurrent impingement or +ve drop arm test (abduct the arm to 90°—ask the patient to lower slowly to side; if unable/pain on lowering, test is +ve). Refer
- **Subacromial impingement** Pain occurs in a limited arc of abduction (60–120°—*painful arc syndrome*) or on internal rotation. If <40y, associated with glenohumeral instability from connective tissue laxity, or labral injury. If older, often due to chronic rotator cuff tendinitis or functional cuff weakness/tear

Investigations X-ray may show calcification of the supraspinatus tendon in acute tendinitis and irregularities/cysts at the humeral greater tuberosity if chronic cuff tendinitis. *Dynamic USS* may demonstrate impingement, tendonitis, and/or rotator cuff tears.

Treatment Rest and NSAIDs, followed by physiotherapy and/or subacromial steroid injection (➔ p. 141). If conservative measures fail refer for specialist management.

Shoulder dislocation Usually due to a fall on arm/shoulder—anterior dislocation is most common. Shoulder contour is lost (flattening of deltoid) and the head of the humerus is seen as an anterior bulge. Axillary nerve may be damaged → absent sensation on a patch below the shoulder. Refer to emergency department for X-ray and reduction. In young patients, ~30% go on to have recurrent dislocations due to labral tear. Dislocation is associated with rotator cuff tear in ~25% of elderly patients.

Recurrent dislocation Usually anterior and follows trauma—but 5% are in teenagers with no trauma but general joint laxity. Refer for specialist physiotherapy and consideration of surgery.

Rupture of the long head of biceps Discomfort in the arm on lifting and a feeling of 'something going'. A lump appears in the body of biceps muscle on elbow flexion. May be associated with other shoulder pathology. *Management*: exclude distal rupture of the tendon at the elbow. Reassure. No treatment necessary.

Acromioclavicular (AC) joint problems Pain on the top of the shoulder or in the suprascapular area suggests a problem with the AC joint or neck. AC joint pain is usually due to trauma or OA—joint tenderness and pain are present on palpation and passive horizontal adduction. *Management*: NSAIDs ± local steroid injection.

Cleido-cranial dysostosis Inherited autosomal dominant condition. Part/all of the clavicle is missing and ossification of the skull is delayed—sutures remain open. Associated with short stature. No treatment.

Elbow problems

History

- **Pain and stiffness** Joint pain is diffuse; pain well localized over the medial or lateral epicondyles may be due to tendinitis
- **Deformity** Swelling? Nodules? Structural deformity?
- **Loss of function** May be limitation of flexion, extension, pronation, and/or supination. This can affect function, e.g. causing difficulty eating (can't get hand to mouth) or with personal care
- **Neurology** Numbness and paraesthesiae distal to the elbow—particularly in the ulnar nerve distribution

Examination

- **Look** Carrying angle (~11° for a ♂; 13° for a ♀). Effusion may be visible either side of the olecranon. A discrete swelling over the olecranon could be RA nodule, gouty tophus, olecranon bursa, or other nodule. Check for muscle wasting
- **Feel** Tenderness? Swelling? Warmth? If indicated test neurology and check pulses distal to the elbow
- **Move** Active and passive movements, Compare both sides. Normal range is from 0° in full extension to 145° in full flexion. Check pronation/supination. Normal range is 75° and 80° respectively

Tennis elbow and golfer's elbow (epicondylitis) Common extensor tendon inflammation at the epicondyle. *Cause*: repeated strain.

- **Tennis elbow**—tenderness over the lateral epicondyle and lateral elbow pain on resisted wrist extension
- **Golfer's elbow**—tenderness over the medial epicondyle and medial elbow pain on resisted wrist pronation

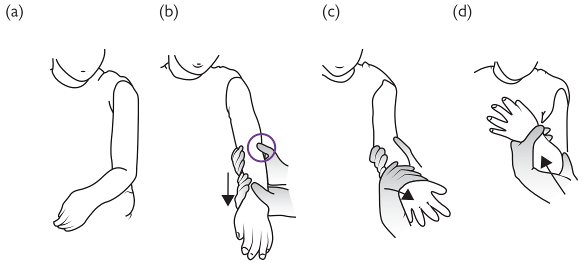
Management Stop trigger movements if possible. Often settles with time ± NSAIDs. Recovery is speeded by local steroid injection (➔ p. 140) but steroid injection may be associated with early recurrence. Physiotherapy may help, as may an epicondylar clasp. Rarely referral for autologous blood injection or surgical release is indicated.

Olecranon bursitis Traumatic bursitis due to repeated pressure on the elbow. Pain and swelling over olecranon. Most settle with conservative treatment (rest, ice, ↓ activity, protective elbow pads). Consider aspiration of fluid from bursa; antibiotics if sepsis suspected. Fluid may reaccumulate—if sepsis has been excluded, consider hydrocortisone injection. Refer if not settling after 2mo or complications.

Ulnar neuritis Narrowing of the ulnar groove (from OA, RA, or post fracture) causes pressure on the ulnar nerve → ulnar neuropathy. Clumsiness with the hand is often the first symptom, then weakness ± wasting of hand muscles innervated by the ulnar nerve and ↓ sensation in the little finger and medial half of the ring finger. Rule out metabolic and autoimmune causes of a mononeuritis and refer for consideration of surgical decompression ± nerve conduction studies if entrapment likely.

Dislocated elbow Usually due to fall on outstretched hand with flexed elbow. Ulna is displaced backwards, elbow is swollen and held in fixed flexion. May have associated fracture. Refer to the emergency department for reduction.

Pulled elbow Common in children <5y (peak age 2–3y). Traction injury to the elbow causes subluxation of the radial head. Often occurs when the child is pulled up suddenly by the hand. The child will not use the arm and generally holds the arm pronated in extension (Figure 14.2a). No other clinical signs. ♂ > ♀. Left arm > right. X-ray is unhelpful. Immediate recovery is seen after reduction (Figure 14.2b–d). If unable to reduce or pain continues after reduction, refer to the emergency department for X-ray and further assessment.



Typical pose:
- Elbow in extension
- Forearm pronated

Step 1:
- Place thumb on prominence of radial head
- Apply gentle longitudinal traction

Step 2:
Supinate the forearm fully

Step 3:
Flex the elbow

Figure 14.2 Technique for reducing a pulled elbow

Further information for patients

Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org

Wrist and hand problems

History

Wrist

- Pain/stiffness Pain is often well localized in the wrist. 5 conditions are associated with point tenderness: de Quervain's disease; old scaphoid fracture; carpometacarpal OA; Kienbock's disease (avascular necrosis of the lunate); tenosynovitis of the extensors. Wrist pain may also be associated with RA, OA, and ganglia. Carpal tunnel syndrome is associated with pain in the hand
- Deformity May be swelling of tendon sheaths or wrist. Bony deformity is a late feature of arthritis or secondary to trauma
- Function Ask about weakness and numbness in the hand

Hand

- Pain/stiffness Pain from the hand is felt in the fingers and/or palm. A diffuse ache may be referred from the neck, shoulder, or mediastinum
- Deformity May occur acutely, e.g. due to tendon rupture or slowly due to bone or joint pathology. The pattern and symmetry of joint involvement can be diagnostic
- Function Good hand function is essential for everyday tasks, e.g. turning keys, doing buttons up, writing. Ask about limitations

Examination

Wrist

- Look Symmetry; swelling; deformity (ulnar deviation, volar subluxation; rheumatoid nodules; ganglia); muscle wasting in forearm/hand
- Feel Temperature; nature of any swellings; tenderness of the radiocarpal, midcarpal, or distal radio-ulnar joint
- Move/measure Range of movement (normal range—extension >75°, flexion >75°, pronation >75° from the vertical, supination >80° from the vertical); crepitation?
- Neurology Check for ulnar and median nerve function

Hand

- Look Posture of the hand; swellings (rheumatoid nodules; Heberden's and Bouchard's nodes; ganglions; tophi); nail signs, e.g. pitting of psoriasis; scars; deformity (mallet finger; swan-neck deformity; boutonnière deformity; Dupuytren's contracture); ulnar deviation. If there is joint disease, note distribution and whether it is symmetrical
- Feel Temperature; condition of the skin, e.g. dryness, sweating; nature of swellings; muscle bulk, e.g. small muscles of the hand; tenderness
- Move/measure Ask the patient to make a fist, spread his/her fingers out, and then test each individual joint. Then test opposition, pinch grip, key grip, palmar grasp of ball, and practical tasks, e.g. picking up a coin

Scaphoid and other hand fractures Table 29.8, ➔ p. 1093

Ganglion Smooth, firm, painless swelling—usually around the wrist. No treatment is needed unless causing local problems. May resolve spontaneously; can be drained (large bore needle)/excised, but often recurs.

⚠ **For all hand injuries** Check for:

Nerve injury Can occur due to trauma or lacerations of the hand or wrist. Examine sensory and motor function. Always ensure no other structures are damaged before suturing skin wounds. Refer all nerve injuries for specialist assessment and management—surgery can improve the outcome considerably in some cases. Intensive hand physiotherapy is important to regain function. Types of nerve injury:

- Neurapraxia Temporary loss of nerve conduction—often caused by pressure causing ischaemia
- Axonotmesis Damage to the nerve fibre but nerve tube is intact—the chance of successful nerve regrowth and a good recovery is high
- Neurotmesis Divided nerve—lack of guidance to the regrowing fibrils gives ↓ chance of a good recovery and a neuroma may develop

Median nerve damage The median nerve controls grasp. Damage causes inability to lift the thumb out of the plane of the palm (abductor pollicis brevis failure) and loss of sensation over the lateral side of the hand.

Ulnar nerve damage Injury distal to the wrist causes a claw hand deformity, loss of abduction/adduction of the fingers and sensory loss over the little finger, and a variable area of the ring finger.

Radial nerve damage The radial nerve opens the fist—injury produces wrist-drop and variable sensory loss including the dorsal aspect of the root of the thumb.

Tendon injury Can occur due to attrition or lacerations of the hand or wrist. Examine hand function. Always ensure no other structures are damaged before suturing skin wounds. Extensor or flexor tendons can be affected. Refer—primary surgical repair is usually the treatment of choice.

Vascular injury Can occur due to trauma/lacerations of the hand or wrist. Check perfusion and temperature of fingers and examine pulses. Ensure no other structures are damaged before suturing skin wounds. Refer all vascular injuries for specialist assessment and management.

Work-related upper limb pain Work related pain in the arm ± wrist, e.g. due to keyboard use. Overuse syndrome. Often termed repetitive strain injury (RSI). Diagnosis of exclusion—no physical signs. Exclude other conditions, e.g. carpal tunnel syndrome (CTS), tennis elbow.

Management Reassure—condition is curable, continue work but avoid the aggravating activity, liaise with work to ensure evaluation of workstation ergonomics. Gradually reintroduce activity. Physiotherapy may help. Explore psychological and work-related issues. A multidisciplinary approach is needed. ❗ Work-related upper limb pain is a notifiable industrial disease.

💣 Existence of RSI has been challenged—rigorous assessment often reveals undiagnosed causes of pain.

Complex regional pain disorder (also known as reflex sympathetic dystrophy or algodystrophy). Pain ± vasomotor changes in a limb → loss of function. Most common in the hand and wrist but may occur in the lower leg/feet. Usually follows trauma—but the trauma may be trivial and signs may appear weeks/months later. *Signs*: pain at rest exacerbated by movement and light touch, swelling, discoloration, temperature changes, abnormal sensitivity, sweating, and loss of function. X-ray may show osteopenia.

Management Physiotherapy improves prognosis if started early; analgesia (NSAIDs, opioids, and/or nerve painkillers). Refer to pain clinic or rheumatology for specialist treatments, e.g. nerve block, spinal cord stimulation, CBT, and/or graded motor imagery.

Tenosynovitis Inflammation of the tendon sheath—often due to unaccustomed activity (e.g. gardening). May affect extensor or flexor tendons. Pain is often worse in the morning. Presents with swelling and tenderness over the tendon sheath and pain on using the tendon. Treat with rest and NSAIDs. If not settling an injection of steroid into the tendon sheath may help. ❗ Notifiable industrial disease if work related

De Quervain's tenosynovitis Tenosynovitis of thumb extensor and abductor tendon sheaths. Pain over radial styloid and on forced adduction/flexion of the thumb. Treat with thumb splint ± local steroid injection. Refer if not settling.

Carpal tunnel syndrome Pain in the radial 3½ digits of the hand ± numbness, pins and needles, and thenar wasting. Due to compression of the median nerve as it passes under the flexor retinaculum. Worse at night. Symptoms are improved by shaking the wrist. *Associations*: pregnancy, hypothyroidism, DM, obesity, and carpal arthritis.

Investigations Phalen's test—hyperflexion of wrist for 1min triggers symptoms; Tinel's test—tapping over the carpal tunnel causes paraesthesiae; request nerve conduction studies if diagnosis is in doubt.

Management GP treatment—night splints may help ± carpal tunnel steroid injection (➔ p. 140). Less likely to help if age >50y or symptoms >10mo. If GP treatment fails, constant paraesthesiae and/or triggering of fingers, refer to orthopaedics for division of the flexor retinaculum.

Kienböck's disease The lunate bone develops patchy necrosis after acute or chronic injury. The patient is usually a young adult complaining of aching and stiffness of 1 wrist. *Examination*: tenderness in the centre of the back of the wrist ± limitation of wrist extension. X-ray is normal at first but later shows ↑ density of the lunate ± deformity. Refer for orthopaedic opinion.

Osteoarthritis in the hand

- Heberden's nodes—swellings of DIP joints. No treatment needed
- Bouchard's nodes—swellings of PIP joints. No treatment needed

First carpometacarpal OA Pain and swelling at the base of the thumb. Thumb becomes stiff. A splint or steroid injection can be helpful. If pain persists surgery (trapeziectomy) may help.

Dupuytren's contracture Palmar fascia contracts so that the fingers (typically the right 5th finger) cannot extend. *Prevalence*: 10% ♂ >65y (more if family history). Less common in women. *Associations*: smoking; alcohol; heavy manual labour; trauma; DM; phenytoin; Peyronie's disease;

AIDS. Often simple reassurance suffices. Ultimately referral for surgery (fasciotomy or fasciectomy) may be needed.

Trigger finger Nodules on the tendon can occur spontaneously and in RA and DM. Most common in ring and middle fingers. The nodule can be palpated moving with the tendon. Pain and triggering (the finger is in fixed flexion and needs to be flicked straight by the other hand) occur because the nodule jams in the tendon sheath. *Management*: local steroid injection or refer for surgical release.

Finger fractures Table 29.8, ➔ p. 1093

Mallet finger The finger tip droops due to avulsion of the extensor tendon attachment to the terminal phalanx. Refer for X-ray. *Management*: a plastic splint which holds the terminal phalanx in extension is worn for 6wk to help union (must not be removed). Arthrodesis may be needed if healing does not occur.

Gamekeeper thumb Forced thumb abduction causes rupture of the ulnar collateral ligament. Can occur on wringing a pheasant's neck—hence the name, or, more commonly, by catching the thumb in the matting on a dry ski slope. The thumb is very painful and pincer grip weak. Refer—open surgical repair is the most effective treatment.

Nail injuries

Avulsed nail Protect the nail bed of an avulsed nail with soft paraffin and gauze, check tetanus status, and give antibiotic prophylaxis (e.g. flucloxacillin 500mg qds for 7d). Partially avulsed nails need removing under ring block to exclude an underlying nail bed injury—the nail is replaced to act as a splint to the nail matrix.

Subungual haematoma A blow to the finger can cause bleeding under the nail—very painful due to pressure build up. Relieve by trephining a hole through the nail using a 19-gauge (cream-coloured) needle (no force required, just twist the needle as it rests vertically on the nail) or a heated point (e.g. of a paper clip or cautery instrument). Of benefit up to 2d after injury.

Polydactyly Extra digits can vary from small fleshy tags to complete duplications. They may be an isolated defect or associated with syndromes. Small fleshy tags are removed in the first few months. For extra digits firmly fixed or involving tendons or joints, surgery is delayed until the child is >1y. Refer to orthopaedics or plastic surgery.

Syndactyly Digits may be joined by a web of skin or more firmly fused. Webbing is usually mild and treatment is for cosmetic reasons if at all. Where digits are fused separation and skin grafting is carried out at ~4y. Refer to plastic surgery.

Hip and pelvis problems

History Pain on walking? Pain at rest? Hip joint pain is usually felt in the groin (Table 14.4). Referred pain is often felt in the knee. Hip disease results in ↓ walking distance, difficulty climbing stairs, and getting out of low chairs.

Examination

- **Look** Watch the patient walk—hip disease → limp or waddling gait
- **Feel** Joint tenderness is just distal to the midpoint of the inguinal ligament
- **Move** Passive movement with the patient lying supine. Check range of movement, pain reproduced on movement? Crepitus?
- **Measure** Hip disease is often associated with shortening of the affected leg—true leg length: anterior superior iliac spine → medial malleolus; apparent leg length: umbilicus → medial malleolus
- **Trendelenburg test** Ask the patient to stand on 1 leg and lift the foot on the contralateral side off the ground. Place your fingers on the anterior superior iliac spines. If the pelvis sags on the unsupported side (+ve Trendelenburg sign) the hip on which the patient is standing is painful or has a weak/mechanically-disadvantaged gluteus medius. ❗ False +ve in 10%

Malignancy Hip and pelvis are common sites for 2° malignancy. Pain is severe and unremitting, day and night. Often accompanied by weight loss. X-ray may show no abnormalities or reveal lytic or sclerotic deposits. Bone scan is diagnostic but may miss myeloma. Depending on clinical circumstances either refer for specialist advice (oncologist, radiotherapist) or to palliative care. Treat with analgesia meanwhile. High risk of pathological fracture.

Osteoarthritis of the hip Major cause of hip pain and disability. Incidence ↑ with age; ♂ ≈ ♀. *Predisposing factors*: past hip disease (e.g. Perthes) or trauma; unequal leg length.

Presentation Pain may be diffuse and felt in hip region, thigh, or knee. Relieved by rest in early stages of disease. *Signs*: ↓ internal rotation and abduction of hip with pain at extremes of movement; antalgic gait; eventually fixed flexion of the hip. *Investigation*: X-ray may confirm diagnosis but is often not needed. There is poor correlation between X-ray changes and pain felt. Perform Oxford Hip Score (Figure 14.3).

Table 14.4 Causes of pain around the hip

Pain	Causes
Buttock pain	PMR, sacroiliitis, vascular insufficiency, referred from back
Groin pain	Hip joint disease (OA, RA, Paget's, osteomalacia), fracture, osteitis pubis, hernia, psoas abscess
Lateral thigh pain	Trochanteric bursitis, referred pain from back, enthesitis (spondyloarthropathies), gluteus medius tear, meralgia paraesthetica, fascia lata syndrome

1. During the past 4 weeks… How would you describe the pain you usually have from your hip?				
None (4)	Very mild (3)	Mild (2)	Moderate (1)	Severe (0)
2. During the past 4 weeks… Have you had any trouble with washing and drying yourself (all over) because of your hip?				
No trouble at all (4)	Very little trouble (3)	Moderate trouble (2)	Extreme difficulty (1)	Impossible to do (0)
3. During the past 4 weeks… Have you had any trouble getting in and out of a car or using public transport because of your hip? (whichever you tend to use)				
No trouble at all (4)	Very little trouble (3)	Moderate trouble (2)	Extreme difficulty (1)	Impossible to do (0)
4. During the past 4 weeks… Have you been able to put on a pair of socks, stockings or tights?				
Yes, easily (4)	With little difficulty (3)	With moderate difficulty (2)	With extreme difficulty(1)	No, impossible (0)
5. During the past 4 weeks… Could you do the household shopping on your own?				
Yes, easily (4)	With little difficulty (3)	With moderate difficulty (2)	With extreme difficulty(1)	No, impossible (0)
6. During the past 4 weeks… For how long have you been able to walk before pain from your hip becomes severe? (with or without a stick)				
No pain/ >30 min (4)	16–30 min (3)	5–15 min (2)	Around the house only (1)	Not at all/pain severe when walking (0)
7. During the past 4 weeks… Have you been able to climb a flight of stairs?				
Yes, easily (4)	With little difficulty (3)	With moderate difficulty (2)	With extreme difficulty(1)	No, impossible (0)
8. During the past 4 weeks… After a meal (sat at a table), how painful has it been for you to stand up from a chair because of your hip?				
Not at all painful (4)	Slightly painful (3)	Moderately painful (2)	Very painful (1)	Unbearable (0)
9. During the past 4 weeks… Have you been limping when walking, because of your hip?				
Rarely/ never (4)	Sometimes, or just at first (3)	Often, not just at first (2)	Most of the time(1)	All of the time (0)
10. During the past 4 weeks… Have you had any sudden, severe pain—'shooting', 'stabbing' or 'spasms'—from the affected hip?				
No days (4)	Only 1 or 2 days (3)	Some days (2)	Most days(1)	Every day (0)
11. During the past 4 weeks… How much has pain from your hip interfered with your usual work (including housework)?				
Not at all (4)	A little bit (3)	Moderately (2)	Greatly (1)	Totally (0)
12. During the past 4 weeks… Have you been troubled by pain from your hip in bed at night?				
No nights (4)	Only 1 or 2 nights (3)	Some nights (2)	Most nights(1)	Every night (0)

Figure 14.3 Oxford Hip Score

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Management of hip osteoarthritis Analgesia (e.g. regular paracetamol, NSAIDs), education, weight ↓, exercise (cycling, swimming), correction of unequal leg length. Walking stick ± shock-absorbing shoe insoles can help. Consider referral for physiotherapy (muscle strengthening exercises may ↓ pain) or, if Oxford Hip Score (Figure 14.3, ➔ p. 463) is ≤20, to orthopaedics for hip injection or replacement.

Total hip replacement >90% achieve good result. Most last >15y. *Post-op care*: risk of dislocation in the first 6wk—advise to avoid crossing legs; take care with transfers; use a walking stick; no driving for 6wk. Physiotherapy is usually arranged via secondary care.

Hip dislocation Occurs in front-seat passengers in car accidents as the knee strikes the dashboard. Reduction under anaesthetic is required.

Greater trochanter pain (trochanteric bursitis) Can mimic ± coexist with hip OA. May be associated with muscle weakness around the hip. *Diagnosis*: point tenderness over the greater trochanter.

Management Consider local steroid injection if trochanteric bursitis is likely. Refer to physiotherapy to strengthen hip musculature to prevent recurrence.

Meralgia paraesthetica Burning/numbness in the upper lateral aspect of the thigh due to compression of the lateral cutaneous nerve of the thigh. *Risk factors*: pregnancy, obesity, DM. *Examination*: extension of the hip or deep palpation just below the anterior superior iliac spine provokes symptoms. *Treatment*: analgesia (including neuropathic painkillers), TENS, ± local steroid injection. Rarely surgical decompression is needed.

Fascia lata syndrome Inflammation of the fascia lata causing pain in the lateral thigh. Often due to overuse or weak musculature. Treatment is with rest ± referral to physiotherapy.

Hip infection Presents with hip pain, ↓ weight, night sweats, and rigors. Be aware of infection in patients with RA, hip prosthesis, or immunocompromise. Refer for investigation. X-rays are often unhelpful—bone scan is non-specific. Admit for USS-guided drainage, bed rest, and IV antibiotics.

Avascular necrosis May present with hip pain. Have a high level of suspicion in patients with risk factors—SLE, sickle cell disease, high alcohol consumption, pregnancy, or corticosteroids. X-ray or bone scan may confirm diagnosis but MRI is most sensitive. Specialist management is needed. Usually progresses to cause OA.

Pubic symphysis dehiscence Painful condition occurring in late pregnancy. May persist after delivery. The pubic symphysis separates resulting in low abdominal pain radiating down both thighs ± low back pain. Pain is constant and worse on movement. It resolves on rest. Examination reveals a soft abdomen and obstetric examination is normal. Advise simple analgesia (e.g. paracetamol). Rest in a semi-recumbent position when in pain. Refer for physiotherapy especially if still a problem in the puerperium. Most resolve spontaneously within several months of delivery. Some persist and need specialist referral

The limping child

- If a child is limping, take it seriously. Look for a problem
- Children find it difficult to localize pain. Pain can be referred from the hip to the knee. Examine the whole limb carefully
- Other causes of referred pain include: spinal pathology, psoas spasm from GI pathology (e.g. appendicitis)
- Limping without pain is uncommon and may be due to undiagnosed developmental dysplasia of the hip—➔ p. 834

Transient synovitis of the hip (irritable hip) The most common reason for limping in childhood. *Peak age*: 2–10y. ♂ > ♀. The child is usually well but complains of pain in the hip or knee and may refuse to weight bear. Often occurs after a viral infection. Cause is unknown. Exclude septic arthritis—refer to orthopaedics. Usually resolves in 7–10d without treatment.

Perthes' disease Pain in the hip or knee, limp, and limited hip movement developing over ~1mo. Due to avascular necrosis of the femoral head. Bilateral in 10%. *Peak age*: 4–7y (range 3–11y). ♂:♀ ≈4:1.

Management If suspected refer for X-ray and to orthopaedics. Treatment is with rest, X-ray surveillance, bracing, and/or surgery depending on severity. Usually heals over 2–3y. Joint damage may cause early arthritis. Risk factors for poor outcome include:

- ♀
- Onset >8y
- Involvement of the whole femoral head
- Pronounced metaphyseal rarefaction
- Lateral displacement of the femoral head

Slipped upper femoral epiphysis The upper femoral epiphysis slips with respect to the femur, usually in a postero-inferior direction. Bilateral in 20%. *Incidence*: 1:100,000. *Peak age*: 10–15y. ♂:♀ ≈3:1. Typically affects obese, underdeveloped children or tall, thin boys.

Presentation Pain at rest in the groin, hip, thigh or referred to the knee; limp and/or pain on movement; ↓ hip movements—particularly abduction and medial rotation. The affected leg may be externally rotated and shortened.

Management Confirm diagnosis on X-ray (include lateral views)—shows backwards and downwards slippage of the epiphysis. Refer to orthopaedics—surgical pinning or reconstructive surgery is needed. Monitoring of the other hip is essential. Complications include: avascular necrosis; coxa vara; early OA; slipped epiphysis on the contralateral side.

Developmental dysplasia of the hip ➔ p. 834

Further information for patients

Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org
Steps Support for patients with lower limb conditions and their families. ☎ 01925 750271 🖱 www.steps-charity.org.uk

Knee problems

History

- **Trauma** History of injury—ask about degree and direction of force
- **Pain/stiffness** Attempt to distinguish well-localized mechanical pain and diffuse inflammatory/degenerative pain
- **Deformity** Swelling? If injury, time of onset of swelling in relation to history (immediate effusion suggests haemarthrosis; post-traumatic effusions appear later). Knock knees or bow legs?
- **Function** Do the Oxford Knee Score (Figure 14.4)

Examination Always compare the 2 knees.

- **Look** Watch the patient walk. Look at the knees while standing—varus/valgus deformity? Ask the patient to lie down. Note quadriceps wasting, scars, skin changes, swelling, and deformity. A space under the knee viewed laterally suggests a fixed flexion deformity. With legs extended, lift both feet off the bed to demonstrate hyperextension
- **Feel** Feel the quadriceps for wasting and palpate the knee for warmth. Check the joint line, collateral ligaments, tibial tubercle, and femoral epicondyles for tenderness. Palpate the popliteal fossa for a Baker's cyst. Check for an effusion. Test for patellofemoral lesions by sliding the patella sideways across the underlying femoral condyles
- **Move** With the patient lying on his/her back, check active and passive range of movement—pain reproduced on movement? Crepitus? Test the medial and lateral collateral ligaments and cruciate ligaments
- **Measure** Quadriceps diameter 18cm up from the joint line in adults

❗ Knee pain can be referred from the hip so examine the hip as well.

Osteoarthritis of the knee Very common; X-ray evidence of OA is even more common. *Treatment*: education; glucosamine; analgesia (paracetamol ± NSAIDs); exercise (refer to physiotherapy). Suggest using a walking stick. Steroid injection can be helpful in some patients. If pain and disability are severe (Oxford Knee Score ≤16), refer to orthopaedics for consideration of total or partial knee replacement. Knee replacement is a very successful procedure resulting in ↓ pain and ↑ mobility. 95% prostheses last >10y.

Infection of the knee joint Most commonly infected joint. *Signs*: hot, red, swollen, painful knee. *Differential diagnosis*: Reiter's disease, gout, pseudogout, traumatic effusion, RA. If infection is suspected refer as an emergency to rheumatology or orthopaedics for investigation. ❗ Do not give antibiotics until the joint has been aspirated.

Non-traumatic knee effusion Common causes: gout, RA, calcium pyrophosphate dehydrate disease (pseudogout), spondyloarthropathies (including reactive arthritis). Consider FBC, ESR, rheumatoid factor, antinuclear antibody, LFTs, bone biochemistry, and thyroid function tests. Drain effusion (or refer to rheumatology to drain) and send fluid for polarized light microscopy (for crystals) and microbiology (?infection).

Management If no infection, inject with long-acting steroid (➔ p. 138). If recurrent and no cause found, refer to rheumatology.

Iliotibial band syndrome ➔ p. 477

1. **During the past 4 weeks…** How would you describe the pain you usually have from your knee?				
None (4)	Very mild (3)	Mild (2)	Moderate (1)	Severe (0)
2. **During the past 4 weeks…** Have you had any trouble with washing and drying yourself (all over) because of your knee?				
No trouble at all (4)	Very little trouble (3)	Moderate trouble (2)	Extreme difficulty (1)	Impossible to do (0)
3. **During the past 4 weeks…** Have you had any trouble getting in and out of a car or using public transport because of your knee? (whichever you tend to use)				
No trouble at all (4)	Very little trouble (3)	Moderate trouble (2)	Extreme difficulty (1)	Impossible to do (0)
4. **During the past 4 weeks…** For how long have you been able to walk before pain from your knee becomes **severe**? (with or without a stick)				
No pain/ >30 min (4)	16–30 min (3)	5–15 min (2)	Around the house only (1)	Not at all/pain severe when walking (0)
5. **During the past 4 weeks…** After a meal (sat at a table), how painful has it been for you to stand up from a chair because of your knee?				
Not at all painful (4)	Slightly painful (3)	Moderately painful (2)	Very painful (1)	Unbearable (0)
6. **During the past 4 weeks…** Have you been limping when walking, because of your knee?				
Rarely/ never (4)	Sometimes, or just at first (3)	Often, not just at first (2)	Most of the time(1)	All of the time (0)
7. **During the past 4 weeks… Could** you kneel down and get up again afterwards?				
Yes, easily (4)	With little difficulty (3)	With moderate difficulty (2)	With extreme difficulty(1)	No, impossible (0)
8. **During the past 4 weeks…** Have you been troubled by pain from your knee in bed at night?				
No nights (4)	Only 1 or 2 nights (3)	Some nights (2)	Most nights(1)	Every night (0)
9. **During the past 4 weeks…** How much has pain from your knee interfered with your usual work (including housework)?				
Not at all (4)	A little bit (3)	Moderately (2)	Greatly (1)	Totally (0)
10. **During the past 4 weeks…** Have you felt that your knee might suddenly 'give way' or let you down?				
Rarely/ never (4)	Sometimes, or just at first (3)	Often, not just at first (2)	Most of the time(1)	All of the time (0)
11. **During the past 4 weeks… Could** you do the household shopping on your own?				
Yes, easily (4)	With little difficulty (3)	With moderate difficulty (2)	With extreme difficulty(1)	No, impossible (0)
12. **During the past 4 weeks… Could** you walk down one flight of stairs?				
Yes, easily (4)	With little difficulty (3)	With moderate difficulty (2)	With extreme difficulty(1)	No, impossible (0)

Figure 14.4 Oxford Knee Score

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Bipartite patella Detected on X-ray. Usually asymptomatic incidental finding but can cause pain due to excessive mobility of a patellar fragment. If troublesome refer for fragment excision.

Patellar dislocation Lateral dislocation of the patella and tearing of the medial capsule/quadriceps can occur due to trauma. More common in young people and if joint hypermobility syndrome. Patient is in pain and unable to flex knee. Refer via A&E or orthopaedics for reduction.

Recurrent subluxation of the patella Medial knee pain + knee 'gives way' due to lateral subluxation of the patella. Most common in girls with valgus knees. *Associations*: familial, hypermobility, high riding patellar. *Signs*: ↑ lateral patellar movement and +ve apprehension test (pain and reflex contraction of quadriceps on lateral patellar pressure). Refer to physiotherapy for vastus medialis exercises. If that is unhelpful, refer to rheumatology to exclude a hereditary connective tissue disorder and/or to orthopaedics for consideration of lateral retinacular release.

Patellar tendinitis Small tear in the patellar tendon causes pain. Most commonly seen in athletes. Differential includes inferior patellar pole enthesitis (spondyloarthropathies), fat-pad syndrome, anterior cartilage lesion, and bursitis. Diagnosis is with USS. Treatment is with rest, NSAIDs, ± steroid injection around (not into) the tendon.

Bursitis Prepatellar bursitis (housemaid's knee) is associated with excess kneeling. Vicar's knee (infrapatellar bursitis) is associated with upright kneeling. Avoid aggravating activity, aspirate ± steroid injection (↓ recurrence). If infected treat with antibiotics ± refer for drainage.

Baker's cyst Popliteal cyst (herniation of joint synovium) can cause swelling and discomfort behind the knee. Usually caused by a degenerative knee. Rupture may result in pain and swelling in the calf mimicking DVT. Treat underlying knee synovitis. Surgical cyst removal may be necessary if persistent problems.

Collateral ligament injury Common in contact sports. Causes knee effusion if severe ± tenderness over the injured ligament. Collateral ligaments provide lateral stability to the knee. Normally there is <5° of movement—if >5° the ligament may be ruptured. Treat with rest, knee support, analgesia. Refer to orthopaedics if rupture is suspected.

Cruciate ligament injury Cruciate ligaments provide anterior/posterior knee stability. Assessment can be difficult.

- Anterior cruciate tears Result from a blow to the back of tibia ± rotation when the foot is fixed on the ground. *Signs*: effusion and +ve draw test (supine with foot fixed and knee at 90°, pull the tibia forward—test is +ve if the tibia moves forward on the femur)
- Posterior cruciate tears Caused, e.g. when the knee hits the dashboard in car accidents. Reverse draw test is +ve (supine with knee at 90°, apply pressure to push the tibia backwards—test is +ve if the tibia moves backward on the femur)

Management Refer to orthopaedics if suspected. Splinting and then physiotherapy helps most (60%) but some require reconstructive surgery—consider urgent referral if keen sportsman.

Chondromalacia patellae Common in teenage girls. Pain on walking up or down stairs or on prolonged sitting. *Signs*: pain on stressing the undersurface of the patella. Arthroscopy (indicated only in severe cases) reveals degenerative cartilage on the posterior surface of the patella. Treat with analgesia + physiotherapy (vastus medialis strengthening ↓ pain in 80%). If persistent, exclude spondyloarthropathy (➔ p. 492) and refer to orthopaedics for arthroscopy.

Osgood–Schlatter disease Seen in athletic teenagers. Pain and tenderness ± swelling over the tibial tubercle. X-rays not required. Avoid aggravating activities. Usually settles over a few months. If not settling refer to orthopaedics or rheumatology for further assessment.

Bow legs and knock knees in children

- **Genu varum (bow legs)** Outward curving of the tibia usually associated with internal tibial torsion. Except in severe cases always resolves spontaneously. Severe cases raise the possibility of rickets or other rare developmental disorders—refer for orthopaedic opinion
- **Genu valgum (knock knees)** Common among 2–4y-olds. Innocent if symmetrical and independent of any other abnormality. Severe, progressive cases suggest rickets—refer for X-ray

Loose bodies in the knee May result in locking of the joint and/or effusion. *Causes*: OA, chip fractures, osteochondritis dissecans, synovial chondromatosis. If problematic refer for removal.

Osteochondritis dissecans Necrosis of articular cartilage and underlying bone. Can cause loose body formation. Cause unknown. Seen in young adults → pain after exercise and intermittent knee swelling ± locking. Predisposes to arthritis. Refer for expert management.

Meniscal lesions Twisting with the knee flexed can cause medial (bucket handle) meniscal tears and adduction with internal rotation can cause lateral cartilage tears. *Symptoms/signs*:

- Locking of the knee—extension is limited due to cartilage fragment lodging between the condyles
- Giving way of the knee
- Tender joint line
- +ve McMurray's test—rotation of the tibia on the femur with flexed knee followed by knee extension causes pain and a click as the trapped cartilage fragment is released. 💣 reliability of this test is debated

Management Refer for MRI ± arthroscopy. Treated by removal of the torn meniscal fragment.

Meniscal cyst Pain + swelling over the joint line due to a meniscal tear. Lateral cysts are more common than medial. The knee may click and give way. Refer for arthroscopy—removal of damaged meniscus relieves pain.

Further information

NICE (2014) Osteoarthritis: care and management. 🖱 www.nice.org.uk/guidance/cg177

Ankle and foot problems

History Trauma; ↑ activity, e.g. walking or running a long way for the patient; feeling of instability; pain/stiffness (relation to weight-bearing; localized/diffuse); deformity (problems getting shoes, shoes wear in odd places or shoes are always uncomfortable); interference with activities.

Examination Compare one foot with the other:

- Look Watch the patient walk normally and on tiptoe. Look at the foot with the patient seated. Check for deformities, the colour of the foot, and any skin/nail changes. Check the shoes for any abnormal patterns of wear (wear is normally under the ball of the foot medially and posterolaterally at the heel)
- Feel Is there any tenderness? Palpate any swellings. Check pulses and skin temperature
- Move Assess active and passive movements of the ankle, subtalar, midtarsal, and toe joints systematically. Check range of movement of joints and pain
- Neurology Check sensation if patient reports any loss of sensation

Ankle, foot, and toe fracture Table 29.8, ➲ p.p. 1093

Achilles tendonitis Inflammation of the Achilles tendon may be related to overuse or a spondyloarthropathy. Presents as a painful local swelling of the tendon. Advise rest. NSAIDs, heel padding, physiotherapy, ± steroid injection may help (never inject into the tendon). If persistent refer to rheumatology.

Ruptured Achilles tendon Presents with a sudden pain in the back of the ankle during activity (felt as a 'kick'). The patient walks with a limp. There is some plantar flexion, but the patient cannot raise the affected heel from the floor when standing on tip toe. A 'gap' can usually be felt in the tendon. Calf squeeze test is −ve (squeezing the calf muscles results in movement of the foot if the Achilles tendon is intact). Refer immediately for consideration of repair. The alternative is immobilization in a splint with the foot plantar flexed.

Pes cavus High foot arches may be idiopathic, due to polio, spina bifida, or other neurological conditions. Toes may claw. Padding under the metatarsal heads relieves pressure. Operative treatment—soft tissue release or arthrodesis—straightens toes. Can lead to tarsal bone OA causing pain—refer for fusion.

Foot drop Patients trip frequently or walk with a high stepping gait. On examination patients are unable to walk on their heels and cannot dorsiflex their foot. Check ankle jerk. *Causes*:

- Common peroneal palsy, e.g. due to trauma—normal ankle jerk
- Sciatica—ankle jerk absent
- L4, L5 root lesion—ankle jerk may be absent
- Peripheral motor neuropathy, e.g. alcoholic—ankle jerk weak or absent
- Distal myopathy—ankle jerk weak or absent
- Motor neurone disease—↑ ankle jerk

Club foot (talipes) Consists of inversion of the foot, adduction of forefoot relative to hindfoot and equinus (plantar flexion).

Positional talipes Moulding deformity seen in neonates. The foot can be passively everted and dorsiflexed to the normal position. Treatment is with physiotherapy. Follow-up to check the deformity is resolving.

True talipes The foot *cannot* be passively everted and dorsiflexed to the normal position. Refer to orthopaedics. Treatment is with physiotherapy, splints, ± surgery.

Flat feet (pes planus) Low medial arch. All babies and toddlers have flat feet. The arch develops after 2–3y of walking. Persistent flat feet may be familial or due to joint laxity. If pain free, foot is mobile, and the patient develops an arch on standing on tiptoe ('flexible' foot), no action is required. If painful may be helped by analgesia, exercises, or insoles. For severe pain, hind foot fusion is an option. Refer if the arch does not restore on tiptoeing ('rigid').

In-toe and out-toe gait

- In-toe Originates in the femur (persistent anteversion of the femoral neck), tibia (tibial torsion), or foot (metatarsus varus). Does not cause pain or affect mobility. Usually resolves by age 5–6y
- Out-toe Common <2y. May be unilateral. Corrects spontaneously

Sever's disease Apophysitis of the heel. *Peak age*: 8–13y. Treated with analgesia, raising the heel of the shoe a little, calf-stretching and avoiding strenuous activities for a few weeks.

Osteochondritis Table 14.5

Syndactyly and polydactyly ➔ p. 461

Table 14.5 Osteochondritis of the foot in children and young adults

	Bone(s) involved	Features	Treatment
Kohler's disease	Navicular bone	Peak age: 3–5y Presents with pain and tenderness over the dorsum of the mid-foot X-ray—small navicular bone of ↑ density	Pain usually resolves with simple analgesia and rest
Freiberg's disease	2nd and 3rd metatarsal heads	Most common in teenagers and young adults. ♀ > ♂ Presents with pain in the foot on walking. The head of the metatarsal is palpable and tender X-ray shows a wide, flat metatarsal	Treatment is usually conservative with cushioning of shoes and simple analgesia. If severe, refer to orthopaedics. Excision of the metatarsal head may relieve pain

Tender heel pad Dull throbbing pain under the heel. Develops a few months after heel trauma. May be due to plantar fasciitis, bursitis, or tendonitis. Treat with rest and heel padding. Refer to physiotherapy—ultrasound treatment can help. Blind steroid injections into the fat pad are not recommended. In persistent cases refer to rheumatology.

Plantar fasciitis/bursitis Common cause of inferior heel pain especially among runners. Pain is worst when taking the first few steps after getting out of bed. Usually unilateral and generally settles in <6wk. Advise shoes with arch support, soft heels, and heel padding (e.g. trainers). Achilles tendon stretching exercises can help; NSAIDs and steroid injection are also helpful. In persistent cases refer to podiatry (for fitting of an insole) ± orthopaedics.

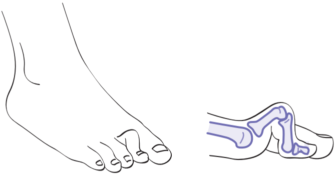
Metatarsalgia (forefoot pain) May be due to synovitis, stress fractures, sesamoid fracture, injury, or ↑ pressure on the metatarsal heads due to mechanical dysfunction (e.g. in RA). Treat with insoles and padding under the metatarsal heads. Surgery may be helpful in RA—discuss with rheumatologist.

Morton's metatarsalgia (interdigital neuroma) Pain due to entrapment of the interdigital nerve between the 3rd/4th metatarsal heads (usually). Gradual onset of sudden attacks of pain or paraesthesia during walking. Refer to orthopaedics. Treatment is with steroid injection and advice regarding footwear. Some need surgical excision of the neuroma.

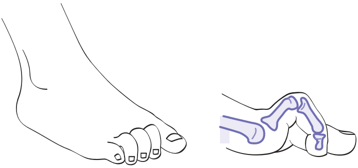
Hammer and claw toes Figure 14.5

- Hammer toes Extended MTP joint, hyperflexed PIP joint, and extended DIP joint. Most common in 2nd toes
- Claw toes Extended MTP joint, flexion at PIP and DIP joints. Due to imbalance of extensors and flexors (e.g. after polio)

If causing pain or difficulty with walking/footwear refer for surgery.



Hammer toe



Claw toes

Figure 14.5 Hammer and claw toes

Achilles tendon stretching exercises

Towel stretch Sit on the floor with your legs stretched out in front of you. Loop a towel around the top of the injured foot. Slowly pull the towel towards you keeping your body straight. Hold for 15–30s then relax—repeat ×10.

Calf/Achilles stretch Stand facing a wall. Place your hands on the wall, chest high. Move the injured heel back and with the foot flat on the floor. Move the other leg forward and slowly lean toward the wall until you feel a gentle stretch through the calf; hold for 15–20s and repeat.

Stair stretch Stand on a step on the balls for your feet, hold the rail or wall for balance. Slowly lower the heel of the injured foot to gently stretch the arch of your foot for 15–20s.

Toe stretch Sit on the floor with knee bent. Pull the toes back on the injured foot until stretch across the arch is felt. Hold for 15–20s and repeat.

Frozen can roll Roll your bare injured foot back and forth from the tip of the toes to the heel over a frozen juice can (not fizzy) or small plastic water bottle. This is a good exercise after activity because it both stretches the plantar fascia and provides cold therapy to the injured area.

Hallux valgus (bunion) Lateral deviation of the big toe at the MTP joint exacerbated by wearing pointed shoes ± high heels. A bunion develops where the MTP joint rubs on footwear. Arthritis at the MTP joint is common. Bunion pads can help but severe deformity requires surgery.

Hallux rigidus Arthritis at 1st MTP joint causes a stiff, painful big toe. Refer severe cases to podiatrist or orthotist for offloading or custom-made rocker bottom foot orthoses. Resistant pain requires surgery.

Ingrowing toe nail Most common in the big toe. Ill-fitting shoes and poor nail cutting predispose to the nail growing into the toe skin → pain. The inflamed tissue is prone to infection. Advise about cutting nails (cut straight with edges beyond the flesh). Refer to podiatry. Treat infection with antibiotics (e.g. flucloxacillin 500mg qds). If recurrent infection, consider referral for surgery (e.g. wedge resection of the nail).

Information for patients

Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org
British Orthopaedic Foot and Ankle Society 🖱 www.bofas.org.uk
Steps Support for patients with lower limb conditions and their families.
☎ 01925 750271 🖱 www.steps-charity.org.uk

Sports medicine

Fitness to perform sporting activities GPs are commonly asked to certify fitness to perform sports. Normally the patient will come with a medical form. If there is a form, request to see it before the medical. If there is no form and you are unsure what to check, telephone the sport's governing body or the event organizer. A fee is payable by the patient.

Many gyms/sports clubs also ask older patients/patients with pre-existing conditions or disabilities to check with their GP before they will sign them on. Assuming that a suitable regime is undertaken, most people can participate. Consider the patient's baseline fitness, check BP and medications, and recommend gradual introduction to new forms of exercise.

❗ Remember—signing a form may result in legal action against you should the patient NOT be fit to undertake an activity. Where possible include a caveat, e.g. 'Based on information available in the medical notes the patient appears to be fit to ... although it is impossible to guarantee this'. If unsure, consult your local LMC/medical indemnity organization.

⚠ *Hypertrophic obstructive cardiomyopathy* Can cause sudden death during sport. It is difficult to exclude on clinical examination—if there is a FH or systolic murmur refer to cardiology before recommending new intense activity.

Benefits of exercise ➔ p. 154

Children and sport

- Exercise is good for children—it stimulates development of the musculoskeletal and cardiovascular systems
- It should be fun and not physically or emotionally over-demanding
- Children are more prone to sports injuries due to continuing growth (bone growth plates are prone to damage) but are more flexible so have ↓ injury rate
- Children's temperature control is not as good as adults
- Equipment must be checked regularly to ensure it fits
- Encourage warm-up and stretching exercises before sport
- Refer children with suspected overuse or sports injuries, which do not recover rapidly with simple analgesia, for specialist assessment

Nutrition Recommend a normal varied diet (➔ p. 148).

- Special circumstances Particular sports have special requirements (e.g. ↑ protein for strength athletes); ↑ muscle glycogen stores before exercise can ↓ fatigue during prolonged heavy exercise, e.g. 'carbohydrate loading'—3–4d of ↑ carbohydrate (8–10g/kg body weight) and a carbohydrate meal 3–4h before competing
- Fluids Sufficient fluid during exercise is vital to good performance and health, especially in hot conditions. Rehydration fluids containing carbohydrate and electrolytes are absorbed faster than plain water
- Supplements e.g. vitamins, minerals, amino acids, carnitine, creatine. A good diet generally supplies sufficient nutrients

Drugs and sport Most regulating bodies have strict codes regarding drug use. Regulations may differ between different sports. Status of a particular medicine may be checked in the Global Drug Reference Online (🖱 www.globaldro.com).

Prohibited classes of drugs

- Stimulants e.g. amphetamine, caffeine (above 12 mcg/mL), ephedrine, certain β_2 agonists (inhaled medication for asthma is allowed)
- Narcotics e.g. morphine, diamorphine, pethidine, methadone (codeine is allowed)
- Anabolic agents e.g. nandrolone, DHEA, testosterone
- Diuretics e.g. furosemide, bendroflumethiazide
- Hormones, hormone antagonists, and related substances e.g. growth hormone, erythropoietin
- Cannabinoids

Classes of drugs subjected to restrictions

- Alcohol and marijuana—restricted in certain sports
- Local anaesthetics—local or intra-articular injection only are allowed (provide written notification of administration)
- Corticosteroids—topical, inhaled, or local/intra-articular injection only are allowed (provide written notification of administration)
- β-blockers—restricted in certain sports

Drugs for pain relief Generally paracetamol, all NSAIDs, and codeine are allowed for pain relief. Stronger opioids and drugs containing caffeine are banned. If in doubt, check on the Global Drug Reference before prescribing.

Anabolic steroid misuse Significant problem in the UK (5% in gyms and fitness clubs). Drugs are often used in complicated regimens at high doses to ↑ lean muscle mass and ↓ body fat. *Side effects include*:

- ↑ cholesterol
- ↑ BP
- Gynaecomastia
- Abnormal LFTs
- Testicular atrophy
- Baldness
- Acne
- Mood changes

❗ Other drugs may be taken in conjunction with anabolic steroids to ↓ these side effects.

> ⚠ Doctors who prescribe or collude in the provision of drugs or treatment with the intention of improperly enhancing an individual's performance in sport risk losing their GMC registration. This does not preclude the provision of any care or treatment where the doctor's intention is to protect or improve the patient's health.

Further information

British Association of Sport and Exercise Medicine 🖱 www.basem.co.uk
Global Drug Reference Online 🖱 www.globaldro.com
MacAuley D (2012) *Oxford Handbook of Sports and Exercise Medicine* (2nd edn). Oxford: Oxford University Press. ISBN: 9780199660155
UKAD Antidoping in Sport. 🖱 www.ukad.org.uk

Management of sporting injuries

Principles of managing sporting injuries

- First aid (**A**irway, **B**reathing, **C**irculation), refer severe injuries to A&E
- RICE
  - **Rest** Relative rest of affected part while continuing other activities to maintain overall fitness
  - **Ice and analgesia** Use immediately after injury (wrap ice in a towel and use for maximum 10min at a time to prevent acute cold injury)
  - **Compression** Taping or strapping can be used to treat (↓ swelling) and also to prevent acute sprains and strains
  - **Elevation** ↓ local swelling and dependent oedema enabling quicker recovery
- **Confirm the diagnosis** Clinical examination, X-ray
- **Early treatment** According to cause. Do not delay
- **Liaise** With sports physician, sports physio, and coach if elite athlete
- **Rehabilitation** Regaining fitness, strength, and flexibility; examine and correct the cause of the injury (e.g. poor technique, equipment)
- **Graded return to activity** Discuss with coach
- **Prevention** Suitable preparation and training (e.g. suitable footwear, warm-up and warm-down exercises, safety equipment) can ↓ likelihood of injuries

Muscle injuries

- **Haematoma** within or between muscles can → dramatic whole limb bruising (due to tracking of blood) and stiffness. Treat with RICE regime, encourage movement in pain-free range
- **Strain** (e.g. hamstring injury) Refer to physiotherapy. A secondary injury is likely if the patient returns to sport too soon

Ligament injuries (sprains)

- **Grade 1** Local tenderness, normal joint movement. Give NSAIDs, support strain, encourage mobilization
- **Grade 2** Slightly abnormal joint movement. More joint protection, NSAIDs, elevate limb, encourage middle of the range movement
- **Grade 3** Abnormal joint movement. Refer to orthopaedics

Groin pain in athletes Consider:

- Conjoint tendon pathology (Gilmore's groin)
- Symphysitis (footballers notably)
- Adductor tendonitis

Liaise with a sports medicine physician or physiotherapist early.

Overuse injuries Incidence is increasing due to increasingly intensive training regimes, especially in young adults—even among amateurs.

- **Causes** Load too great for conditions, poor technique or posture, faulty or poor quality equipment
- **Types of injury** Stress fractures, joint tenderness or effusion, ligament and tendon strains, muscle stiffness

- **Management** Rest, NSAIDs, physiotherapy, improved training regime
- **Prevention** Recognize and correct poor posture or technique, check equipment is appropriate and fits, warm up and stretching before exercise, gradually ↑ intensity and duration of training

Shin splints Exercise-related shin pain may be due to a stress fracture of the tibia, compartment syndrome, or periostitis. Fractures are not always seen on X-ray—bone scan is more sensitive and shows periostitis. Treat with rest and analgesia. Consider referral to sports physiotherapist.

Iliotibial band syndrome Pain due to inflammation of the synovium under the iliotibial tract from rubbing of the tract on the lateral femoral condyle. Seen in runners. Treat with rest, NSAIDs, specialist physiotherapy ± steroid injection.

Over-training syndrome Poor performance, fatigue, heavy muscles, and depression due to excessive sports training or competing without sufficient rest. Usually diagnosed from history. Exclude other causes of fatigue (➔ p. 502). Manage with rest, reassurance, and alteration of training programme.

'Scrumpox' (herpes gladiatorum) Herpes simplex virus is very contagious and outbreaks among sporting teams are common, e.g. spread by close contact and facial stubble grazes while scrumming. *Treatment*: aciclovir (cream or tablets) and exclusion of infected players. Impetigo, erysipelas, and tinea barbae can be transmitted in the same way.

Environmental factors

- **Heat cramps** Painful spasm of heavily exercised muscles (calves and feet)—due to salt depletion. *Treatment*: rest, massage of affected muscle, and fluid and salt replacement (e.g. Dioralyte®)
- **Heat stroke/exhaustion** Exercising in excessive heat → salt and water depletion, dehydration, and metabolite accumulation. *Signs*: headache, nausea, confusion, incoordination, cramps, weakness, dizziness, and malaise. Eventually thermoregulatory mechanisms fail → seizures and coma. *Signs*: flushing, sweating, and dehydration. Temperature may be normal (mild cases) or ↑. *Treatment*: Rest, fluid and salt replacement (e.g. Dioralyte®). Admission for IV fluids and supportive measures in severe cases
- **Hypothermia** Ensure appropriate clothing and limit time in the cold. *Signs*: behaviour change, incoordination, clouding of consciousness. *Treatment*: remove from cold environment, wrap in blankets (including the head), and transfer to hospital. Do not use direct heat
- **Frost bite** Freezing of the peripheries (usually feet, hands, ears, or nose). Tissues become hard, insensitive, and white. *Treatment*: gentle rewarming, Refer if significant dead tissue. Debridement is usually delayed to allow natural recovery
- **Diving** Decompression illness is due to rapid ascent causing nitrogen dissolved in blood to form gas bubbles. Usually <1–36h after surfacing. *Presentation*: deep muscle aches, joint pains, skin pain, paraesthesia, itching and burning, retrosternal pain, cough and breathlessness, neurological symptoms. Refer suspected cases urgently to A&E

Bone disorders

Osteogenesis imperfecta Inherited condition with autosomal dominant inheritance (rarely recessive). Several types but all have an underlying problem with collagen metabolism resulting in fragile bones that break easily. Other features include lax joints, thin skin, blue sclerae, hypoplastic teeth, and deafness. Presentation varies according to severity. May be obvious at birth or present early with fractures. Less severe cases present later and may be mistaken for NAI. Mild cases may not present until adolescence with thin bones on X-ray. Treatment is supportive.

Osteopetrosis (marble bone disease) Inherited condition with autosomal dominant or recessive inheritance. Dominant form presents in childhood with fractures, osteomyelitis ± facial paralysis. Recessive form is more severe causing bone marrow failure and death. Bone marrow transplantation has been tried but is of limited success.

Paget's disease of bone Abnormal osteoclast activity causes accelerated, disorganized bone remodelling. Affects 1–2% of UK adults; 15% have a FH. ♂: ♀ ≈3:1. Most common in the elderly—only a minority are symptomatic. Affects just one bone in 1 in 3 cases.

Presentation Pain—dull ache aggravated by weight bearing, often remains at rest; deformity—bowing of weight-bearing bones especially tibia (sabre), femur, and forearm—usually asymmetrical; frontal bossing of the forehead; distinctive changes on X-ray; ↑ bone-specific alk phos; normal Ca^{2+}, PO_4, and PTH.

Management Refer to rheumatology. Give analgesia. Oral/IV bisphosphonates ↓ pain and long-term complications. *Complications*: pathological fracture; OA of adjacent joints; high-output CCF; hydrocephalus and/or cranial nerve compression → neurological symptoms, e.g. deafness; spinal stenosis; bone sarcoma (rare: 0.1–1.15%).

Osteomyelitis Infection of bone. May spread from abscesses or follow surgery. Often no primary site is found. More common in those with DM, sickle cell disease, impaired immunity, and/or poor living standards. *Organisms involved: S. aureus*, streptococci, *E. coli, Salmonella, Proteus*, and *Pseudomonas* species, TB. Presents with pain, unwillingness to move affected part, warmth, effusions in neighbouring joints, fever, and malaise. Blood cultures are +ve in 60%; ↑ ESR/CRP; ↑ WCC.

Management Refer suspected cases for same-day orthopaedic opinion. Diagnosis is confirmed with imaging, e.g. MRI or bone scan (X-ray changes can take days to appear). *Treatment*: is with IV then PO antibiotics (≥6wk) and surgery to drain abscesses. *Complications*: septic arthritis, pathological fracture, deformity of growing bone, chronic infection.

Chronic osteomyelitis Occurs after delayed/inadequate treatment of acute osteomyelitis. *Signs*: pain, fever, and discharge of pus from sinuses. Follows a relapsing/remitting course over years. Needs specialist management.

⚠ Referral guidelines for suspected sarcoma[N]

Refer for immediate X-ray Any patient with suspected spontaneous fracture. If the X-ray:

- Indicates possible bone cancer, refer urgently
- Is normal but symptoms persist, follow-up and/or request repeat X-ray, bone function tests, or referral

Refer urgently If a patient presents with a palpable lump that is:

- >5 cm in diameter
- Deep to fascia, fixed, or immobile
- Increasing in size
- Painful
- A recurrence after previous excision

❗ If a patient has HIV, consider Kaposi sarcoma and make an urgent referral if suspected.

Urgently investigate Increasing, unexplained, or persistent bone pain or tenderness, particularly pain at rest (and especially if not in the joint), or an unexplained limp. In older people metastases, myeloma, or lymphoma, as well as sarcoma, should be considered.

Sarcoma Cancer of the bone or connective tissue. ~2300 patients/y are diagnosed with sarcoma in the UK, and it causes ~1000 deaths. There are 2 peaks of incidence—one in teenagers, and another in old age. 5 types of sarcoma account for >80% of tumours.

Osteosarcoma and the Ewing's family of tumours Present with aching bone pain, swelling, ± pathological fracture. If X-ray is normal but symptoms persist, consider checking bone function tests, re-X-raying, discussing the patient with a specialist or referral. Treatment involves surgery and chemotherapy. Overall 5y survival is 50–80%.

Adult soft tissue sarcoma of limb or trunk Usually presents with a palpable lump. The most common tumours are leiomyosarcoma, liposarcoma, and synovial sarcoma. Treated with surgery ± radiotherapy (high-grade tumours). Chemotherapy is reserved for palliation.

Kaposi sarcoma ➔ p. 722

Intra-abdominal sarcoma Usually presents late. Often arises in the retroperitoneum. If possible, surgery is the main treatment. Local relapse is common and often not responsive to cytotoxic therapy.

Rhabdomyosarcoma Originates from striated muscle. Presents usually in children <2y with a lump. Responds to intensive multimodal therapy; outlook is generally good (>60% long-term survival).

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Patient information and support

Brittle Bone Society ☎ 01382 204446 🖱 www.brittlebone.org
Paget's Association ☎ 0161 799 4646 🖱 www.paget.org.uk
Sarcoma UK ☎ 0808 801 0401 🖱 www.sarcoma.org.uk

Rickets and osteomalacia

Vitamin D deficiency causes rickets in children and osteomalacia in adults. The body needs ~10 micrograms of vitamin D per day to maintain healthy bones. The body makes its own vitamin D when sunlight falls on the skin in the summer months but a diet with adequate vitamin D is needed to maintain the supply in the winter—especially for people who do not get out or for cultural or religious reasons are completely shielded from the sun by their clothing.

Clinical features of rickets

- Bone pain/tenderness: arms, legs, spine, pelvis
- Skeletal deformity: bow legs, pigeon chest (forward projection of the sternum), rachitic rosary (enlarged ends of ribs), asymmetrical/odd-shaped skull due to soft skull bones, spinal deformity (kyphosis, scoliosis), pelvic deformities
- Pathological fracture
- Dental deformities—delayed formation of teeth, holes in enamel, ↑ cavities
- Muscular problems—progressive weakness, ↓ muscle tone, muscle cramps
- Impaired growth → short stature (can be permanent)

Clinical features of osteomalacia

- Bone pain—diffuse, particularly in hips
- Muscle weakness
- Pathological fractures
- Low calcium → perioral numbness, numbness of extremities, hand and feet spasms, and/or arrhythmias

Causes and management

Dietary deficiency (<30nmol/L) Particularly in children with pigmented skin in Northern climes. Give vitamin D and Ca^{2+} supplements.

Age-related deficiency (<30nmol/L) Vitamin D metabolism deteriorates with age and many >80y are deficient. Consider giving vitamin D (800IU/d) to all elderly >80y.

Secondary rickets/osteomalacia Due to other disease, e.g. malabsorption, liver disease, renal tubular disorders, or chronic renal failure. Treat underlying cause and supplement Ca^{2+} and vitamin D as needed.

Vitamin D-dependent rickets Rare autosomal recessive inherited disorder resulting in an enzyme deficit in the metabolism of vitamin D. Refer for specialist care. Treated with vitamin D and Ca^{2+} supplements.

Hypophosphataemic rickets (vitamin D-resistant rickets) X-linked dominant trait resulting in ↓ proximal renal tubular resorption of phosphate. Parathyroid hormone and vitamin D levels are normal. Specialist management is needed. Treatment is with phosphate replacement ± calcitriol.

Dietary sources of calcium and vitamin D Tables 14.6 and 14.7

Table 14.6 Approximate vitamin D content of common foods

Food	Serving	Vitamin D (mcg)[a]
Margarine	10g (½ oz)	0.8
Eggs	1 size 3	1.1
Cheese	60g (2oz)	0.2
Milk	0.15l (¼ pint)	0.05
Butter	10g (½ oz)	0.1
Fortified cereals	30g (1oz)	0.5
Herring	100g (3½ oz)	16.5
Mackerel	100g (3½ oz)	8
Sardines	100g (3½ oz)	7.5
Tinned tuna	100g (3½ oz)	4
Tinned salmon	100g (3½ oz)	12.5
Kipper	100g (3½ oz)	13.5

[a] *Recommended daily intakes*: birth to 50y, 5mcg; 50–70y, 10mcg; >70y, 15mcg.

Table 14.7 Approximate calcium content of common foods

Food	Serving	Calcium (mg)[a]
Whole milk	0.2l (⅓ pint)	220
Semi-skimmed milk	0.2l (⅓ pint)	230
Hard cheese	30g (1oz)	190
Cottage cheese	115g (4oz)	80
Low-fat yoghurt	150g (5oz)	225
Sardines (including bones)	60g (2oz)	310
Brown or white bread	3 large slices	100
Wholemeal bread	3 large slices	55
Baked beans	115g (4oz)	60
Boiled cabbage	115g (4oz	40

[a] *Recommended daily intakes*: birth to 6mo, 210mg; 7mo–1y, 270mg; 1–3y, 500mg; 4–8y, 800mg; 9–18y, 1300mg; 19–50y, 1000mg; >50y, 1200mg.

Patient information and support

Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org

Osteoporosis

Lifetime risk of osteoporotic fracture is 1:3 in ♀ and 1:5 in ♂ (>200,000 fractures/y in the UK). The main morbidity and financial costs of osteoporosis relate to hip fracture where incidence ↑ steeply >70y. Treatment aims to prevent fracture.

Definitions

- **T-score** Compares bone mineral density (BMD) of the subject with the young adult mean (age 30y)
- **Osteoporosis** Defined as BMD >2.5 standard deviations (SD) below the young adult mean (i.e. T score of <−2.5). There is 2–3× ↑ relative risk of fracture for each SD ↓ in BMD
- **Osteopenia** Diagnosed if T-score is between −1 and −2.5
- **Z-score** Compares BMD of the subject to an age-matched normal control, i.e. measures whether BMD is normal for the patient's age. Cannot be used to diagnose osteoporosis or osteopenia, but may be useful in young patients to predict osteoporosis risk for the future

Causes Osteoporosis may be 1° or 2° to other medical conditions:

- **Endocrine** Hypogonadism (e.g. premature menopause, anorexia, androgen blockade, taking aromatase inhibitors), hyperthyroidism, hyperparathyroidism, hyperprolactinaemia, Cushing's disease, type 1 DM
- **GI** Coeliac disease or other causes of malabsorption, inflammatory bowel disease, chronic liver disease, chronic pancreatitis
- **Rheumatological** RA, other inflammatory arthropathies
- **Other** Immobility, multiple myeloma, haemoglobinopathy, systemic mastocytosis, CF, COPD, CKD, homocystinuria

Fragility fracture Fracture sustained falling from ≤ standing height—includes vertebral collapse (may not be as a result of a fall). Previous fracture is a risk for future fracture. Common fractures:

- **Hip** Associated with ↑ mortality
- **Wrist** Colles' fracture
- **Osteoporotic vertebral collapse** Causes pain, ↓ height, and kyphosis. Pain can take 3–6mo to settle and requires strong analgesia. Calcitonin is useful for pain relief for 3mo after vertebral fracture if other analgesics are ineffective

Predicting fracture risk[N] 2 validated fracture risk prediction tools are available: FRAX® (available from 🖱 www.shef.ac.uk/FRAX) and Qfracture® (available from 🖱 www.qfracture.org). Qfracture does not require BMD measurement; FRAX can be performed without BMD measurement. Both provide information on 10y probability of hip or other osteoporotic fracture. Case-finding and further actions—Figure 14.6.

Bone mineral density (BMD) measurement X-rays cannot be used to measure BMD, but are useful if vertebral fracture or metastases is suspected. Hip and lumbar spine BMD is measured using dual energy X-ray absorptiometry (DXA). Do not request without prior use of a risk prediction tool, e.g. FRAX® (without BMD measurement) or Qfracture®.

Glucocorticoid use Steroid use is a risk factor for osteoporosis. Minimize steroid dose. For patients taking oral/high-dose inhaled steroids for >3mo or frequent courses of steroids, in addition:

- Add bone protection agent (e.g. bisphosphonate) for patients >65y or with history of fragility fracture *or*
- Refer patients <65y without history of fragility fracture for DXA scan and add a bone protection agent if T score is ≤−1.5

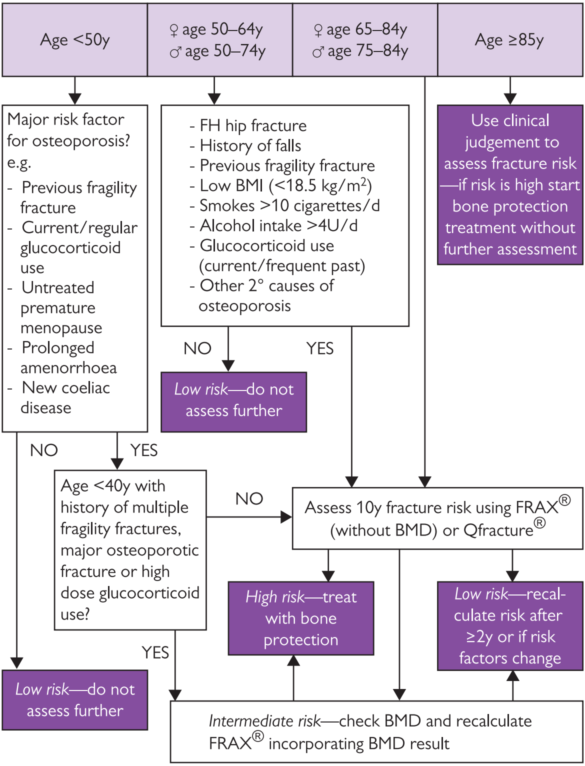

Age <50y
♀ age 50–64y
♂ age 50–74y
♀ age 65–84y
♂ age 75–84y
Age ≥85y
Major risk factor for osteoporosis? e.g.
- Previous fragility fracture
- Current/regular glucocorticoid use
- Untreated premature menopause
- Prolonged amenorrhoea
- New coeliac disease
- FH hip fracture
- History of falls
- Previous fragility fracture
- Low BMI (<18.5 kg/m²)
- Smokes >10 cigarettes/d
- Alcohol intake >4U/d
- Glucocorticoid use (current/frequent past)
- Other 2° causes of osteoporosis
Use clinical judgement to assess fracture risk —if risk is high start bone protection treatment without further assessment
NO
YES
Low risk—do not assess further
NO
YES
Age <40y with history of multiple fragility fractures, major osteoporotic fracture or high dose glucocorticoid use?
NO
Assess 10y fracture risk using FRAX® (without BMD) or Qfracture®
High risk—treat with bone protection
Low risk—recalculate risk after ≥2y or if risk factors change
YES
Low risk—do not assess further
Intermediate risk—check BMD and recalculate FRAX® incorporating BMD result

Figure 14.6 Use of fracture risk prediction tools

Further information

National Osteoporosis Guideline Group Clinical guideline for the prevention and treatment of osteoporosis. 🖱 www.shef.ac.uk/NOGG

NICE (2012, updated 2017) Assessing the risk of fragility fracture. 🖱 www.nice.org.uk/guidance/cg146

Treatment options for osteoporosis

Lifestyle advice Provide to all at-risk patients.

- Adequate nutrition
  - Maintain body weight so BMI >19kg/m^2 and adequate intake of calcium and vitamin D (Tables 14.6 and 14.7, ➔ p. 481)
  - Give Ca^{2+} and/or vitamin D supplements to postmenopausal women with dietary deficiency; also consider if on long-term steroids, >80y, housebound, or institutionalized
- Regular exercise Weight-bearing activity >30min/d ↓ fracture rate
- Stop smoking Women who stop smoking pre-menopause have a 25% ↓ fracture rate post-menopause
- ↓ alcohol consumption to <14U/wk (♂ and ♀)

Osteopenia If T-score of between −1 and −2.5, provide lifestyle advice. Repeat DXA scan in ~2y.

Bisphosphonates e.g. alendronic acid 70mg once weekly. ↓ bone loss and fracture rate. Mainstay of treatment for osteoporosis. Avoid if severe CKD or woman of child-bearing age (possible teratogenic effects).

Instructions for use Take on an empty stomach first thing in the morning, ≥30min before food/other medication; take in an upright position washed down with plenty of water; sit upright for 30min after taking.

Osteonecrosis of the jaw Rare complication of bisphosphonate therapy (IV > po preparations). Causes non-healing gum lesions. Treatment is with surgical excision of the affected bone. Risk of osteonecrosis ↑ after dental work—advise patients to have a dental check-up and any necessary dental work done before starting bisphosphonate treatment, and report any oral symptoms when on treatment to their dentist.

Atypical femoral fracture Prolonged bisphosphonate treatment >5y/ oversuppression of bone turnover and ↑ bone fragility. Acute sub-trochanteric or mid-shaft femoral fractures are most common. 💣 A 'drug holiday' of 1–5y has been proposed for low-risk patients after 5y use—follow local guidance.

Denosumab 60mg sc every 6mo. Monoclonal antibody that ↓ osteoclast activation and ↓ bone resorption.

- For postmenopausal osteoporosis when bisphosphonates are contraindicated/not tolerated and severe osteoporosis^{N}
- Can be used for women with severe CKD; correct hypocalcaemia before starting treatment. May cause osteonecrosis of the jaw

Raloxifene 60mg od. Selective oestrogen receptor modulator (SERM).

- Use if previous fragility fracture, bisphosphonates are not tolerated or contraindicated, or there is an unsatisfactory response with bisphosphonates (further fracture and/or ↓ in BMD after ≤1y treatment). Not recommended for primary prevention of osteoporotic fracture^{N}
- Avoid if past history of DVT/PE, cholestasis, endometrial cancer, or undiagnosed vaginal bleeding

HRT (➔ p. 688). Postpones postmenopausal bone loss and ↓ fractures. Optimum duration of use is uncertain (>5–7y) but benefit disappears <5y after stopping. ↑ in breast cancer and cardiovascular risk limits use.

⚠ *NICE guidance (2015)*

- Premature menopause HRT or COC is recommended for the prevention of osteoporosis until women reach natural menopause age
- >51y HRT should *not* be considered first-line therapy for long-term prevention of osteoporosis. HRT remains an option where other therapies are contraindicated, cannot be tolerated, or if there is a lack of response; risks and benefits should be carefully assessed

Teriparatide Third line for postmenopausal women and second line for men with past history of fragility fracture if other treatments are not tolerated/ineffective and specific T-score and clinical criteria are met. Given by daily injection. Maximum duration of use is 18mo. Consider referral for consultant initiation if other treatment options are exhausted.

Osteoporosis in men Currently only bisphosphonates and teriparatide are recommended for treatment of osteoporosis in men.

Monitoring There is no consensus about duration of treatment for osteoporosis or monitoring of BMD during treatment. Circumstances in which repeat DXA scanning might be necessary include:

- Fragility fracture on treatment
- If considering a change in treatment
- When considering restarting therapy after a drug holiday

Referral Consider referral to an appropriate specialist if (*U* = urgent referral; *R* = routine referral):

- Another cause for fragility fracture is suspected (e.g. metastasis)—*U*
- Fragility fracture on treatment—*R*
- Unusual presentation of osteoporosis, e.g. premenopausal woman—*R*
- For consideration of treatment with IV bisphosphonate, denosumab, or teriparatide—*R*

Further information

National Osteoporosis Guideline Group Clinical guideline for the prevention and treatment of osteoporosis. 🖱 www.shef.ac.uk/NOGG

NICE (2008, updated 2018) Alendronate, etidronate, risedronate, raloxifene, strontium ranelate and teriparatide for the primary and secondary prevention of osteoporotic fragility fractures in postmenopausal women. 🖱 www.nice.org.uk/guidance/ta161

NICE (2010) Osteoporotic fractures—denosumab. 🖱 www.nice.org.uk/guidance/ta204

NICE (2015) Menopause: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng23

Osteoarthritis

Osteoarthritis (OA) is the most important cause of locomotor disability. It used to be considered 'wear and tear' of the bone/cartilage of synovial joints but is now recognized as a metabolically active process involving the whole joint—i.e. cartilage, bone, synovium, capsule, and muscle.

The main reason for patients seeking medical help is pain. Level of pain and disability are greatly influenced by the patient's personality, anxiety, depression, and activity, and often do not correlate well with clinical signs.

Risk factors ↑ age (uncommon <45y); ♀ > ♂; ↑ in black and Asian populations; genetic predisposition; obesity; abnormal mechanical loading of joint, e.g. instability; poor muscle function; post-meniscectomy; certain occupations, e.g. farming.

Symptoms and signs Joint pain ± stiffness, synovial thickening, deformity, effusion, crepitus, muscle weakness/wasting, and ↓ function. Most commonly affects hip, knee, and base of thumb. Typically exacerbations last weeks to months. Nodal OA, with swelling of the distal interphalangeal joints (Heberden's nodes) has a familial tendency.

Investigations X-rays may show ↓ joint space, cysts and sclerosis in subchondral bone, and osteophytes. OA is common and may be a coincidental finding. Exclude other causes of pain, e.g. check FBC and ESR if inflammatory arthritis is suspected (normal or mildly ↑ in OA—ESR >30mm/h suggests RA or psoriatic arthritis).

Management of osteoarthritis in primary care Employ a holistic approach. Assess effect of OA on the patient's functioning, quality of life, occupation, mood, relationships, and leisure activities. Formulate a management plan with the patient that includes self-management strategies, effects of co-morbidities and regular review.

Information and advice Give information and advice on all relevant aspects of osteoarthritis and its management. Arthritis Research UK produces a range of leaflets for patients. Use the whole multidisciplinary team, e.g. refer to:

- Physiotherapist for advice on exercises, strapping, and splints
- OT for aids
- Chiropodist for foot care and insoles
- Social worker for advice on disability benefits and housing
- Orthopaedics for surgery if significant disability/night pain

↓ load on the joint Weight reduction can ↓ symptoms and may ↓ progression in knee OA. Using a walking stick in the opposite hand to the affected hip and cushioned insoles/shoes (e.g. trainers) can also help.

Exercise and improving muscle strength ↓ pain and disability, e.g. walking (for OA knee), swimming (for OA back and hip but may make neck worse), cycling (for OA hip and OA knee—but may worsen patellofemoral OA). Refer to physiotherapy for advice on exercises especially isometric exercises for the less mobile.

Pain control

- Use non-pharmacological methods first (activity, exercise, weight ↓, footwear modification, walking stick, TENS, local heat/cold treatments)

- Regular paracetamol (1g qds) is first-line drug treatment for all OA and/or topical NSAIDs for knee/hand OA only. Topical NSAIDs have less side effects than oral NSAIDs and are more acceptable to patients
- Use opioids, oral NSAIDs, or COX2 inhibitors as second-line agents in addition to, or instead of paracetamol. Use the lowest effective dose for the shortest possible time. Co-prescribe a proton pump inhibitor (e.g. omeprazole 20mg od) with NSAIDs if taking for >1wk
- Low-dose antidepressants, e.g. amitriptyline 10–75mg nocte (unlicensed) are a useful adjunct especially for pain causing sleep disturbance
- Capsaicin cream can also be helpful for knee/hand OA[N]

Aspiration of joint effusions and joint injections Can help in exacerbations. Some patients respond well to long-acting steroid injections—it may be worth considering a trial of a single treatment. Hyaluronic acid knee injections are not recommended by NICE.

Complementary therapies ~60% of sufferers from OA are thought to use CAM, e.g. copper bracelets, acupuncture, food supplements, dietary manipulation. There is good evidence chiropractic/osteopathy can be helpful for back pain, but otherwise evidence of effectiveness is scanty. Advise patients to find a reputable practitioner with accredited training who is a member of a recognized professional body and carries professional indemnity insurance.

💣 *Other drugs/supplements*

- Glucosamine It is controversial whether glucosamine modifies OA progression. It is available OTC but not recommended by NICE
- Strontium ranelate ↓ progression of OA, ↓ pain, and ↑ mobility[R]. Place in OA management is yet to be determined

Psychological factors Have a major impact on the disability from OA. Education about the disease, and emphasis that it is not progressive in most people, is important. Seek depression and anxiety with screening tools—➔ p. 173. Treat as needed.

Refer

- To rheumatology To confirm diagnosis if coexistent psoriasis (psoriatic arthritis mimics OA and can be missed by radiologists); rule out 2° causes of OA (e.g. pseudogout, haemochromatosis) if young OA or odd distribution; if joint injection is thought worthwhile but you lack expertise or confidence to do it
- To orthopaedics If symptoms are severe for joint replacement. Refer as an emergency if you suspect joint sepsis

Further information

NICE Osteoarthritis: care and management. 🖱 www.nice.org.uk/guidance/cg177

Information and support for patients

Arthritis Care ☎ 0808 800 4050 🖱 www.arthritiscare.org.uk
Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org

Rheumatoid arthritis

Rheumatoid arthritis (RA) is the most common disorder of connective tissue affecting ~1% of the UK population. It is an immunological disease, triggered by environmental factors, in patients with genetic predisposition. Disease course is variable with exacerbations and remissions.

⚠ Refer all suspected cases of rheumatoid arthritis to rheumatology—early treatment with disease-modifying drugs can significantly alter disease progression. Refer urgently[N] if:

- Small joints of the hands/feet are affected
- >1 joint is affected
- There has been a delay of ≥3mo between onset of symptoms and seeking medical advice

Presentation

- Can present at any age—most common in middle age. ♀:♂ ≈3:1
- Variable onset—often gradual but may be acute
- Usually starts with symmetrical small joint involvement—i.e. pain, stiffness, swelling, and functional loss (especially in the hands)—joint damage and deformity occur later
- Irreversible damage occurs early if untreated and can → deformity and joint instability
- Other presentations—monoarthritis; migratory (palindromic) arthritis; PMR-like illness; systemic illness of malaise, pain, and stiffness

Symptoms and signs Predominantly peripheral joints are affected—symmetrical joint pain, effusions, soft tissue swelling, early morning stiffness. Progression to joint destruction and deformity. Tendons may rupture. Specific features—Table 14.8.

Differential diagnosis Diagnosis may not be easy—consider:

- Psoriatic arthritis
- Nodal OA
- SLE (especially in ♀ <50y)
- Bilateral carpal tunnel syndrome
- Other connective tissue disorders
- Polymyalgia rheumatica if >50y

Investigations

- Check FBC (normochromic normocytic or hypochromic, microcytic anaemia), ESR, and/or CRP (↑). May have ↑ platelets, ↓ WCC
- Rheumatoid factor and anti-CCP antibodies are +ve in the majority. A minority have a +ve ANA titre
- X-rays—normal, periarticular osteoporosis or soft tissue swelling in the early stages; later—loss of joint space, erosions, and joint destruction

Management A multidisciplinary team approach is ideal, e.g. GP, medical and surgical teams, physiotherapist, podiatrist, OT, nurse specialist, and social worker.

Screening for depression ➔ p. 173

General support Provision of information about the disease, treatments, and support available (including equipment and help with everyday activities, self-help and carers groups, disabled parking badges, financial support—➔ p. 108).

Physical therapy Exercises, splints, appliances, and strapping help to keep joints mobile, ↓ pain, and preserve function.

Table 14.8 Specific features of rheumatoid arthritis

Hands	• Ulnar deviation of the fingers • 'z' deformity of the thumb • Swan neck (hyperextended PIP and flexed DIP joints) and boutonnière (flexed PIP and extended MCP joints, hyperextended DIP joint) deformities of the fingers (Figure 14.7) • ↓ grip strength and ↓ hand function causes disability
Legs and feet	• Subluxation of the metatarsal heads in feet and claw toes → pain on walking • Baker's cyst (➔ p. 468) at the knee may rupture mimicking DVT
Spine	Especially cervical spine—causing neck pain, cervical subluxation, and atlanto-axial instability leading to a risk of cord compression. X-rays are required prior to general anaesthesia
Non-articular features	Common. Weight ↓, fever, malaise. • *Rheumatoid nodules* (especially extensor surfaces of forearms) • *Vasculitis*—digital infarction, skin ulcers, mononeuritis • *Eye*—Sjögren's syndrome, episcleritis, scleritis • *Lungs*—pleural effusions, fibrosing alveolitis, nodules • *Heart*—pericarditis, mitral valve disease, conduction defects • *Skin*—palmar erythema, vasculitis, rashes • *Neurological*—nerve entrapment, e.g. carpal tunnel syndrome, mononeuritis, and peripheral neuropathy • *Felty's syndrome*—combination of RA, splenomegaly, and leucopenia. Occurs in patients with long-standing RA. Recurrent infections are common. Hypersplenism → anaemia and thrombocytopenia. Associated with lymphadenopathy, pigmentation, and persistent skin ulcers. Splenectomy may improve the neutropenia

C-reactive protein (CRP) Acute phase protein that ↑ ≤6h after an acute event. Follows clinical state more rapidly than ESR (➔ p. 636). Not ↑ by SLE, leukaemia, UC, pregnancy, OA, anaemia, polycythaemia, or heart failure. Highest levels are seen in bacterial infections (>10mg/L).

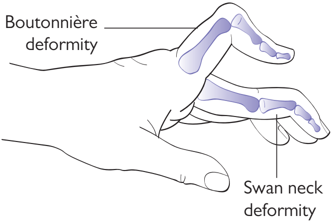


Figure 14.7 Boutonnière and swan neck deformities of the fingers

Information and support for patients

Arthritis Care ☎ 0808 800 4050 🖱 www.arthritiscare.org.uk
Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org

Medication

NSAIDs and simple analgesics e.g. regular paracetamol. Provide symptomatic relief but do not alter the course of disease. Patients' response to NSAIDs is individual—start with the least gastric toxic, e.g. ibuprofen 200–400mg tds and alter as necessary, e.g. to naproxen 500mg bd. If the patient has a history of indigestion/gastric problems, consider adding gastric protection, e.g. PPI, or, if there is no history of CVD, using a COX2 inhibitor, e.g. celecoxib 100mg bd.

Corticosteroids Intra-articular injections of steroids (e.g. triamcinolone) can settle localized flares (e.g. knee or shoulder) and can be used up to 3×/y in any particular joint. Depot IM injections or IV infusions (pulses) can also help settle an acute flare but offer short-term benefits with the risk of systemic side effects. Daily low-dose oral steroids help symptoms and there is some evidence that they can modify disease progression, but concerns about adverse side effects have limited use.

Disease-modifying drugs (DMARDs)

- Methotrexate
- Sulfasalazine
- Penicillamine
- Gold
- Azathioprine
- Leflunomide
- Hydroxychloroquine
- Ciclosporin
- Cyclophosphamide
- Biologic therapies, e.g. rituximab, infliximab, etanercept, adalimumab

Use only under consultant supervision. ↓ disease progression by modifying the immune response and inflammation. Used individually or in combination, they are now started very early in the disease (i.e. first 3–6 mo)—hence the need for early referral. DMARDs can take several months to show any effect. Before starting check baseline U&E, Cr, eGFR, LFTs, FBC and urinalysis. Side effects and monitoring—Table 14.9.

⚠ *Results requiring action*

- Total WBC <3.5 × 10^9/L
- Neutrophils <2 × 10^9/L
- Platelets <150 × 10^9/L
- LFTs (ALT/AST) >2× baseline
- Persistent proteinuria (>1+ ×2) or haematuria

Discuss with rheumatologist ± stop medication.

Surgery Aims to relieve pain and improve function. Consideration of the risks, benefits, and the most appropriate timing of surgery is vital. Common procedures include: joint fusion, replacement, and excision; tendon transfer and repair; and nerve decompression.

Complications of RA Physical disability, depression, osteoporosis, ↑ infections, lymphoma, cardiovascular disease, amyloidosis (10%), side effects of treatment.

Further information

British Society for Rheumatology (2017) Guideline for the prescription and monitoring of non-biological DMARD drugs. 🖱 https://www.rheumatology.org.uk/practice-quality/guidelines

NICE (2018) Rheumatoid arthritis in adults. 🖱 www.nice.org.uk/guidance/ng100

Primary Care Rheumatology Society 🖱 www.pcrsociety.org.uk

Table 14.9 Specific disease-modifying drugs—side effects and monitoring

Drug	Routine monitoring	Side effects to monitor
Methotrexate 7.5–25mg weekly It is common practice to give folate 5mg the day after methotrexate (i.e. weekly) as well	FBC, U&E, eGFR, and LFT weekly until dose and monitoring are stable. Then monthly for at least 1y Frequency of monitoring may be ↓ by specialist if disease/dose stable after 1y CXR within 1y of start of treatment. Check baseline lung function if lung disease	Ask to report symptoms/signs of infection—especially sore throat If severe respiratory symptoms <6mo after starting, refer to A&E If MCV >105fL check vitamin B_{12}/folate
❗ Advise patients NOT to self-medicate with aspirin or ibuprofen. Avoid alcohol		
Sulfasalazine 1g bd/tds maintenance	FBC and LFT monthly for first 3mo. Then every 3mo Urgent FBC if intercurrent illness during initiation If stable after a year, frequency of monitoring may be ↓ by specialist	Rash (1%) Nausea/diarrhoea—often transient Bone marrow suppression in 1–2% in the first months If MCV >105fL check vitamin B_{12}/folate
Intramuscular gold (Myocrisin®) 50mg monthly	FBC and urinalysis at the time of each injection CXR within 1y of start of treatment	*Ask patients to report*: Symptoms/signs of infection—especially sore throat Bleeding/bruising Breathlessness/cough Mouth ulcers/metallic taste *or* rashes
Penicillamine 500–750mg/d maintenance	FBC, urinalysis 2-weekly for 3mo and 1wk after any ↑ dose. Then monthly	Altered taste (can be ignored), rash
Azathioprine 1.5–2.5mg/kg/d maintenance	FBC and LFT weekly for 6wk, then every 2wk until dose/monitoring stable for 6wk. Then monthly	GI side effects, rash, bone marrow suppression Avoid live vaccines
⚠ If allopurinol is co-prescribed, ↓ dose to 25% of the original		
Ciclosporin 1.25mg/kg bd maintenance	FBC and LFT monthly until dose/monitoring stable for 3mo, then every 3mo U&E, Cr/eGFR every 2wk until dose stable for 3mo, then monthly Lipids 6-monthly	Rash, gum soreness, hirsutism, renal failure/↑ Cr (if ↑ by >30% from baseline, withhold and discuss with rheumatologist), ↑ BP Monitor BP
Hydroxychloroquine 200–400mg/d maintenance	Baseline eye check and annual check of visual symptoms and visual acuity	Rash, GI effects, ocular side effects (rare)
Leflunomide 10–20mg/d maintenance	FBC and LFT monthly for 6mo then, if stable every 2mo	Rash, GI, ↑ BP, ↑ ALT Check weight and BP at each review

⚠ Before starting check baseline U&E, Cr, eGFR, LFTs, FBC, and urinalysis.

The spondyloarthropathies

A group of inflammatory rheumatic diseases characterized by predominant involvement of axial and peripheral joints and entheses (areas where tendons, ligaments, or joint capsules attach to bone). Includes:

- Ankylosing spondylitis
- Psoriatic arthritis
- Reactive arthritis and Reiter's syndrome
- Behçet's disease
- Arthritis that accompanies inflammatory bowel disease
- Whipple's disease (➲ p. 379)

Sacroiliitis and spondylitis occur with all of them, and they are all associated with the HLA B27 genotype.

Ankylosing spondylitis (AS) Prevalence 1 in 2000. ♂:♀ ≈2½:1. 95% HLA B27 +ve—prevalence in a population mirrors the frequency of the HLA B27 genotype. Risk of developing AS if HLA B27 +ve ≈1:3.

Presentation Typically presents with morning back pain/stiffness in a young man. Progressive spinal fusion (ankylosis) leads to ↓ spinal movement, spinal kyphosis, sacroiliac (SI) joint fusion, neck hyperextension and neck rotation. *Other features*:

- ↓ chest expansion
- Chest pain
- Hip and knee arthritis
- Plantar fasciitis and other enthesopathies
- Iritis
- Crohn's or UC
- Heart disease—carditis, aortic regurgitation, conduction defects
- Osteoporosis
- Psoriasiform rashes

Tests

- Blood FBC—normochromic or microcytic hypochromic anaemia, ↑ ESR (may be normal), rheumatoid factor is usually −ve
- X-ray Initial signs are widening of the SI joints and marginal sclerosis—later SI joint fusion and a 'bamboo spine' (vertebral squaring/fusion)

Management Aims to ↓ inflammation, pain, and stiffness; alleviate systemic symptoms, e.g. fatigue; and slow or stop long-term progression of the disease. Exercise helps back pain. NSAIDs (e.g. naproxen 500mg bd) also help pain. Refer to a rheumatologist early for confirmation of diagnosis, education, disease-modifying drugs (➲ p. 490), and advice on appropriate exercise regimes to maintain mobility.

Psoriatic arthritis Inflammatory arthritis associated with psoriasis (~40% psoriasis patients. ♂ = ♀). 75% patients have a pre-existing history of psoriasis before the arthropathy; in 15% the rash appears simultaneously with the joint symptoms; in 10% the arthritis precedes the skin changes. Presentation is variable. Patterns include:

- Distal arthritis DIP joint swelling of hands/feet, nail dystrophy, ± flexion deformity. Sausage-shaped fingers are characteristic of psoriatic arthritis affecting the hand

- **Rheumatoid-like polyarthropathy** similar to rheumatoid arthritis (➔ p. 488) but less symmetrical and rheumatoid factor is −ve
- **Mutilans** Associated with severe psoriasis. Erosions in small bones of hands/feet ± spine. Bones dissolve → progressive deformity
- **Ankylosing spondylitis/sacroiliitis** Usually HLA B27 +ve

Investigations WBC—usually ↑; ESR/CRP—usually ↑; rheumatoid factor–ve; X-ray appearances can be diagnostic.

Management Education; physiotherapy; NSAIDs. Refer to rheumatology for confirmation of diagnosis, advice on management, and disease-modifying drugs (➔ p. 490). Medication, e.g. methotrexate, may improve both skin and musculoskeletal symptoms.

Reactive arthritis Often asymmetrical aseptic arthritis in ≥1 joint. Occurs 2–6wk after bacterial infection elsewhere—e.g. gastroenteritis (*Salmonella, Campylobacter*), GU infection (*Chlamydia, Gonorrhoea*). ↑ incidence in HLA B27 +ve individuals.

Management NSAIDs, physiotherapy, and steroid joint injections. Recovery usually occurs within months. A minority develop chronic arthritis requiring disease-modifying drugs. Refer to rheumatology.

Reiter's syndrome Polyarthropathy, urethritis, iritis, and a psoriasiform rash. Affects men with HLA B27 genotype. Commonly follows GU or bowel infection. Joint and eye changes are often severe. Refer for specialist management.

Behçet's disease Multi-organ disease of unknown cause (although thought to be infective). ♂:♀ ≈2:1. *Clinical picture* (only some features): arthritis; ocular symptoms and signs—pain, ↓ vision, floaters, iritis; scarring painful ulceration of mouth and/or scrotum; colitis; meningoencephalitis. Refer to GUM clinic, ophthalmologist or general physician depending on symptom cluster. Treatment is usually with steroids ± azathioprine or ciclosporin. Topical steroids may be useful for ulcers.

Enteropathic spondyloarthropathy Oligoarticular or polyarticular arthritis linked to inflammatory bowel disease. Presentation is variable and includes: sacroiliitis, plantar fasciitis, inflammatory spinal pains, and other enthesitides (insertional ligament/tendon inflammation). Arthritis may evolve and relapse/remit independently of bowel disease.

Management NSAIDs may help joint pain but aggravate bowel disease. Refer to rheumatology for confirmation of diagnosis and advice on management and disease-modifying drugs.

Information and support for patients

Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org
National Ankylosing Spondylitis Society (NASS) ☎ 020 87411515 🖱 www.nass.co.uk
Psoriasis and Psoriatic Arthritis Alliance (PAPAA) ☎ 01923 672837 🖱 www.papaa.org

Crystal-induced arthritis

Hyperuricaemia ↑ serum uric acid. *Causes*:

- **Drugs** Cytotoxics; diuretics; ethambutol
- **↑ cell turnover** e.g. lymphoma; leukaemia; psoriasis; haemolysis
- **↓ excretion** Primary gout; CKD; hyperparathyroidism
- **Disorders of purine synthesis** e.g. Lesch–Nyhan syndrome

Gout^{G} Intermittent attacks of acute joint pain due to deposition of uric acid crystals. *Prevalence*: 3–8/1000. ↑ with age; ♂:♀ ≈5:1. *Risk factors*:

- FH
- Obesity
- Excess alcohol
- High-purine diet (Table 14.10)
- Acute infection
- Surgery
- CKD
- Plaque psoriasis
- Polycythaemia
- Leukaemia
- Diuretics
- Cytotoxics

❗ Untreated gout is associated with ↑ risk of death, CVD, and CKD.

Presentation

- **Acute gout** Painful swollen joint (distal joints, e.g. big toe, feet, and ankles, most commonly); red skin which may peel ± fever. Can be polyarticular—especially in elderly ♀. May mimic septic arthritis
- **Chronic gout** Recurrent acute attacks, tophi (urate deposits) in pinna, tendons, and joints ± joint damage

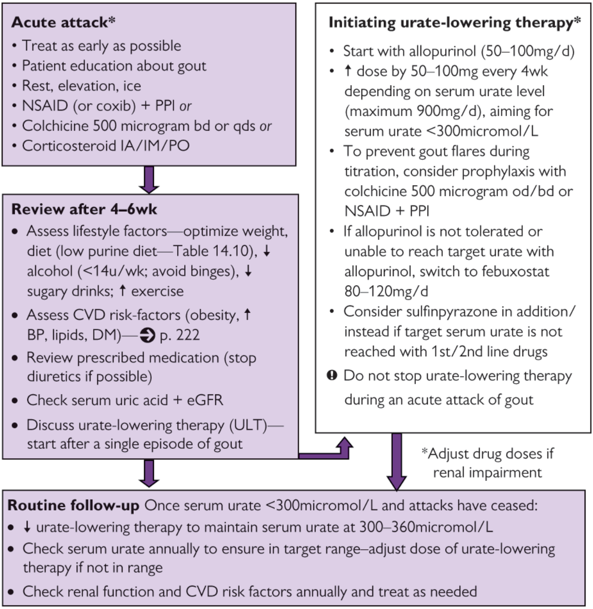
Investigation Usually a clinical diagnosis. If investigations are required, consider: blood (↑ WCC; ↑ ESR; normal or ↑ uric acid); microscopy of synovial fluid (sodium monourate crystals on polarized light microscopy); X-ray (soft tissue swelling unless severe disease when erosive pattern).

Management Figure 14.8. Refer to rheumatology if serum urate cannot be controlled with drugs available in primary care or if symptoms are not settling despite treatment.

Calcium pyrophosphate deposition disease (CPPD) Also known as *pseudogout*. Inflammatory arthritis due to deposition of pyrophosphate crystals. Associated with OA, hyperparathyroidism, and haemochromatosis. Attacks are less severe than gout and may be difficult to differentiate from other types of arthritis. Knee, wrist, and shoulder are most commonly affected. Acute attacks can be triggered by intercurrent illness. Chondrocalcinosis may be seen on X-ray (calcification of articular cartilage). Presence of joint crystals confirms diagnosis.

Management Treat acute attacks like acute gout. A chronic form also occurs—frequently erosive. Refer to rheumatology for confirmation of diagnosis and advice on management and disease-modifying drugs.

⚠ **Septic arthritis** The most important differential diagnosis for acute gout. It is most common in children <5y old and most commonly affects the hip or knee, but septic arthritis can occur at any age and affect any joint. The patient is usually systemically unwell and holds the affected joint completely still. The joint may be swollen, hot, and tender. This is an orthopaedic emergency—if suspected admit. Treatment is with IV antibiotics ± surgical washout of the joint.



Acute attack*

- Treat as early as possible
- Patient education about gout
- Rest, elevation, ice
- NSAID (or coxib) + PPI *or*
- Colchicine 500 microgram bd or qds *or*
- Corticosteroid IA/IM/PO

Review after 4–6wk

- Assess lifestyle factors—optimize weight, diet (low purine diet—Table 14.10), ↓ alcohol (<14u/wk; avoid binges), ↓ sugary drinks; ↑ exercise
- Assess CVD risk-factors (obesity, ↑ BP, lipids, DM)—➔ p. 222
- Review prescribed medication (stop diuretics if possible)
- Check serum uric acid + eGFR
- Discuss urate-lowering therapy (ULT)—start after a single episode of gout

Initiating urate-lowering therapy*

- Start with allopurinol (50–100mg/d)
- ↑ dose by 50–100mg every 4wk depending on serum urate level (maximum 900mg/d), aiming for serum urate <300micromol/L
- To prevent gout flares during titration, consider prophylaxis with colchicine 500 microgram od/bd or NSAID + PPI
- If allopurinol is not tolerated or unable to reach target urate with allopurinol, switch to febuxostat 80–120mg/d
- Consider sulfinpyrazone in addition/instead if target serum urate is not reached with 1st/2nd line drugs

❗ Do not stop urate-lowering therapy during an acute attack of gout

*Adjust drug doses if renal impairment

Routine follow-up Once serum urate <300micromol/L and attacks have ceased:

- ↓ urate-lowering therapy to maintain serum urate at 300–360micromol/L
- Check serum urate annually to ensure in target range–adjust dose of urate-lowering therapy if not in range
- Check renal function and CVD risk factors annually and treat as needed

Figure 14.8 Management of gout

Table 14.10 Purine content of common foods

High purine	Moderate purine	Low purine
Oily fish, e.g. mackerel, herring, sardines, trout, whitebait Seafood, e.g. lobster, crab, prawns, mussels, fish roe, caviar Game, e.g. venison, pheasant, rabbit Meat/yeast extracts, e.g. beer, Marmite®, Bovril® Offal, e.g. liver, kidneys, heart, sweetbreads	Most meat, e.g. beef, pork, lamb Most poultry, e.g. chicken, turkey Mushrooms/Quorn™ Processed legumes, e.g. baked beans, kidney beans, soya products Some vegetables, e.g. asparagus, cauliflower, spinach Wholegrains, e.g. bran, wholemeal bread	Dairy products, e.g. milk, cheese, butter, yoghurt Eggs Bread/cereal (not wholegrain) Pasta/noodles Most fruit/vegetables

Further information

British Society for Rheumatology (2017) Guideline for the management of gout. https://www.rheumatology.org.uk/practice-quality/guidelines

Connective tissue diseases

Group of overlapping diseases that affect many organs, and are associated with fever, malaise, chronic (often relapsing/remitting) course, and response to steroids. Often difficult to diagnose.

Systemic lupus erythematosus (SLE) Autoimmune disease with prevalence of 1 in 3000; ♀:♂ ≈9:1. ↑ in Afro-Caribbeans and Asians. Onset 15–40y. Presentation—Table 14.11; there *must* be multisystem involvement.

Investigations Check an autoimmune profile—95% are ANA (anti-nuclear antibody) +ve. *Other immunological abnormalities*—↑ double-stranded DNA, RhF +ve (40%), ↓ complement (C3, C4). *FBC*: ↓ Hb, ↓ WCC, ↑ ESR.

Management Refer to rheumatology. Use NSAIDs for symptom control. Sunscreens protect skin (ACBS). Steroids are the mainstay of treatment of acute flares (always discuss with a rheumatologist). Hydroxychloroquine can improve skin and joint symptoms. Cyclophosphamide, methotrexate, and ciclosporin are also used.

⚠ Sulfonamides and hormonal contraceptives/HRT may worsen SLE.

Drug-induced lupus Occurs with:
- Minocycline
- Isoniazid
- Hydralazine
- Procainamide
- Chlorpromazine
- Sulfasalazine
- Losartan
- Anticonvulsants

Remits slowly when the drug is stopped but steroids may be needed.

Discoid lupus erythematosus (LE) ♀:♂ ≈2:1. ≥1 well-defined, red, round/oval plaques on the face, scalp, or hands. Scarring may → scalp alopecia and skin hypopigmentation. Internal involvement is not a feature. Confirm with lesion biopsy. Investigate with an autoimmune profile as for

Table 14.11 Presentation of SLE

System	% of patients	Presenting complaints	
Joints	95	• Arthritis • Arthralgia	• Myalgia • Tenosynovitis
Skin	80	• Photosensitivity • Facial 'butterfly' rash • Vasculitic rash	• Hair loss • Urticaria • Discoid lesions
Lungs	50	• Pleurisy • Pneumonitis	• Pleural effusion • Fibrosing alveolitis
Kidney	50	• Proteinuria • ↑ BP	• Glomerulonephritis • Renal failure
Heart	40	• Pericarditis	• Endocarditis
CNS	15	• Depression • Psychosis • Infarction	• Fits • Cranial nerve lesions
Blood	95	• Anaemia (very common) • Thrombocytopenia	• Splenomegaly
Fatigue	95		

SLE. Treat with potent topical steroids and sunscreen. Remission occurs in 40%. 5% develop SLE.

Antiphospholipid syndrome ↑ clotting tendency occurring with SLE or alone. Associated with thrombosis, stroke, migraine, miscarriage, myelitis, MI, and multi-infarct dementia. If suspected, start aspirin 150mg od and refer to rheumatology. May need anticoagulation.

Sjögren's syndrome

- **Primary Sjögren's syndrome** Under-recognized cause of fatigue and dryness of skin/mucous membranes (may present with dyspareunia). Often presents with nodal OA. Long-term, associated with lymphoma. Autoimmune profile is characteristic
- **Secondary Sjögren's syndrome** Association of any connective tissue disease (50% have RA) with keratoconjunctivitis sicca (↓ lacrimation → dry eyes) or xerostomia (↓ salivation → dry mouth)

Management Refer to rheumatology. Provide information/support. Use artificial tears for dry eyes. Xerostomia may respond to frequent cool drinks, artificial saliva sprays, e.g. Glandosane®, or sugar-free gum. Inform dentist of the diagnosis. Rashes may respond to antimalarials.

Raynaud's syndrome Intermittent digital ischaemia precipitated by cold or emotion. Fingers ache and change colour: pale → blue → red on rewarming. Usually presents <25y of age and is idiopathic. *Prevalence*: 3–20%; ♀ > ♂; often abates at the menopause; 5% develop autoimmune rheumatic disease—mainly scleroderma and SLE.

Differential diagnosis

- Other rheumatology conditions—scleroderma; SLE; RA
- Haematology conditions—leukaemia; polycythaemia; thrombocytosis; cold agglutinins; monoclonal gammopathy; mixed cryoglobulinaemia
- Drugs, e.g. β-blockers
- Thoracic outlet obstruction
- Smoking/arteriosclerosis
- Trauma, e.g. use of vibrating tools

Management Keep warm—socks/gloves/hats in cold weather, hand warmers, stay inside. Avoid drugs that ↑ symptoms, e.g. β-blockers. Stop smoking. Nifedipine 10–20mg tds, amlodipine 5mg od, or fluoxetine 20mg od (unlicensed) may help. If associated/severe symptoms refer to rheumatology (urgently if critical ischaemia, e.g. ulceration/infarcts on fingers).

Systemic sclerosis Spectrum of disorders causing fibrosis and skin tightening (scleroderma). Raynaud's is usually present, ± ↑ BP, lung fibrosis, GI symptoms, telangiectasia, polyarthritis, and myopathy. Provide education/support. Treat symptoms. Early specialist referral is vital. CREST (*C*alcinosis of subcutaneous tissues; *R*aynaud's; o*E*sophageal motility problems; *S*clerodactyly and *T*elangiectasia) has better prognosis.

Further information and support for patients

British Sjögren's Association ☎ 0121 478 1133 🖱 www.bssa.uk.net
British Society for Rheumatology (2017) Guideline for the management of adults with SLE. 🖱 https://www.rheumatology.org.uk/practice-quality/guidelines
Lupus UK ☎ 01708 731251 🖱 www.lupusuk.org.uk
Scleroderma and Raynaud's UK ☎ 0800 3112756 🖱 www.sruk.co.uk

Polymyalgia and giant cell arteritis

Polymyalgia rheumatica (PMR) and giant cell (or temporal) arteritis (GCA) are 2 clinical syndromes that are part of the same spectrum. *Key features*:

- Both PMR and GCA affect the elderly (rare <50y); ♀:♂ ≈3:1
- 50% of patients with GCA also have PMR; 15% with PMR have GCA
- Both conditions usually respond rapidly and dramatically to corticosteroids

Presentation Diagnosis is clinical. Both PMR and GCA may present with malaise, anorexia, fever, night sweats, weight ↓, and depression. Check ESR/CRP on presentation.

Diagnosis of PMR A person may be regarded as having PMR if the following criteria are present[G]:

- Age >50y; duration >2wk
- Bilateral shoulder or pelvic girdle aching, or both
- Morning stiffness duration of >45min
- Evidence of acute phase response, i.e. ↑ ESR (usually >30mm/h) or ↑ CRP). ❗ Diagnosis can be made without ↑ inflammatory markers if classical clinical picture and rapid response to steroid treatment

Diagnosis of GCA A person may be regarded as having GCA if ≥3 of the following criteria are met[G]:

- Age ≥50y
- New headache—unilateral throbbing headache, facial pain, scalp tenderness, e.g. on brushing hair, and/or jaw claudication (↑ likelihood of visual symptoms)
- Temporal artery abnormality (tenderness, thickening, ↓ pulsation)
- ↑ ESR >50mm/h (or ↑ CRP)
- Abnormal temporal artery biopsy compatible with GCA

❗ Visual symptoms (amaurosis fugax, diplopia, or sudden loss of vision) are early complications of GCA and may be the presenting feature.

Differential diagnosis

PMR

- Inflammatory arthritis, e.g. RA
- Connective tissue disease/vasculitis, e.g. SLE
- OA
- Septic arthritis
- Shoulder disease
- Neoplasia, e.g. myeloma
- Occult sepsis, e.g. endocarditis
- Inflammatory myopathy
- Fibromyalgia
- Endocrinopathy/metabolic bone disease

GCA

- Herpes zoster
- Migraine
- Intracranial pathology
- Other causes of acute vision loss, e.g. amaurosis fugax
- Cluster headache.
- Cervical spondylosis or other cervical spine disease
- Sinus/ear disease
- TMJ pain
- Connective tissue disease/vasculitis

Further investigation Intended to exclude other diagnoses:

- Full blood count—normocytic anaemia may be seen in PMR/GCA
- U&E/eGFR
- Liver function tests
- Bone profile
- TFTs
- Creatine kinase
- Dipstick urinalysis

- Protein electrophoresis (also consider urinary Bence Jones protein)
- Rheumatoid factor (consider ANA and anti-CCP antibodies too)
- Consider CXR and/or hip/pelvis/shoulder/cervical spine X-ray

Further management of PMR If typical symptoms/signs management in primary care is appropriate.

- Start prednisolone 15mg od—there should be a rapid response (≥70% ↓ of symptoms in <1wk); if not, question the diagnosis. ESR/CRP should return to normal in <4wk
- Continue prednisolone 15mg od for 3wk then ↓ dose to 12.5mg od for 3wk, then 10mg od for 4–6wk, then by 1mg every 4–8wk. Tailor steroid regimen to the patient—↑ time at each dose may be needed
- If there is relapse of symptoms, go back to the previous higher dosage
- At the start of treatment give osteoporosis prophylaxis (➔ p. 484) and supply with a steroid card (➔ p. 276)
- Usually 1–2y of treatment is needed. The need for ongoing treatment >2y should prompt referral for specialist assessment

Referral for rheumatology assessment

- 'Red flag' features: prominent systemic features, weight ↓, night pain, neurological signs (urgent referral)
- Symptoms of GCA
- Younger patient <60y
- Chronic onset (over >2mo)
- Lack of shoulder involvement
- Lack of inflammatory stiffness
- Normal or very high ESR/CRP
- Peripheral arthritis or other features of CTD/muscle disease
- Treatment dilemmas, e.g. incomplete/non-response to steroids; treatment required >2y

Further management of GCA

- Corticosteroids ↓ vascular complications, particularly blindness, and relieve symptoms (70% improvement in <1wk); prescribe prednisolone 40–60mg daily to start immediately a diagnosis of GCA is suspected
- At the start of treatment give osteoporosis prophylaxis (➔ p. 484) and supply with a steroid card (➔ p. 276)
- Consider starting aspirin 75mg od
- Refer urgently to ophthalmology or rheumatology depending on local referral pathways for temporal artery biopsy and ongoing management (same day if visual symptoms)

❗ Temporal artery biopsy may be −ve even in cases of GCA due to skip lesions. Do not withhold treatment while waiting for biopsy—but if the patient has had steroids ≥2wk +ve biopsy is less likely.

Further information

British Society for Rheumatology (2009) Management of polymyalgia rheumatica. 🖱 https://www.rheumatology.org.uk/practice-quality/guidelines

British Society for Rheumatology (Due 2019) Management of giant cell arteritis. 🖱 https://www.rheumatology.org.uk/practice-quality/guidelines

Vasculitis

Characterized by inflammation within or around blood vessels ± necrosis. Severity depends on size and site of vessels affected. Systemic vasculitis can be life threatening. *Causes*:

- Idiopathic (50%)
- Connective tissue disease (e.g. RA, SLE)
- Infection (e.g. rheumatic fever, infective endocarditis, Lyme's disease)
- Drugs (e.g. NSAIDs, antibiotics)
- Neoplasia (e.g. lymphoma, leukaemia)

Presentation Variable—may be confined to the skin or systemic involving joints, kidneys, lungs, gut, and nervous system.

- **Skin signs** Palpable purpura (often painful)—usually on lower legs/buttocks
- **Systemic effects** Fever, night sweats, malaise, weight ↓, myalgia, and arthralgia may occur in all types of vasculitis

Conditions Table 14.12—many are rare.

Table 14.12 Vasculitic conditions

Condition	Features	Management
Erythema nodosum	➔ p. 568	➔ p. 568
Henoch–Schönlein purpura (HSP)	More common in children than adults; ♂ > ♀ Presents with a purpuric rash over buttocks and extensor surfaces Platelet count is normal Often follows a respiratory infection Other features: urticaria, nephritis, joint pains, abdominal pain (may mimic acute abdomen)	Refer to paediatrics for confirmation of diagnosis Most recover fully without treatment over a few months
Polyarteritis nodosa (PAN)	Uncommon in the UK. ♂:♀ ≈4:1. Peak incidence in middle age Multi-system necrotizing vasculitis → aneurysms of medium-sized arteries Presents with: tender subcutaneous nodules along the line of arteries, coronary arteritis, ↑ BP, mononeuritis multiplex, renal failure, and GI symptoms Sometimes associated with hepatitis B	Refer to rheumatology for angiography to confirm diagnosis, and for advice on management Treatment is with control of ↑ BP, high-dose steroids and cyclophosphamide
Eosinophilic granulomatosis with polyangiitis (formerly Churg–Strauss syndrome)	Associated with asthma Affects coronary, pulmonary, cerebral, and splanchnic circulations Skin manifestations and mononeuritis can also occur Diagnosis is based on clinical features and biopsy	Refer for specialist treatment with high-dose prednisolone ± cyclophosphamide Avoid leukotriene receptor agonist drugs for control of asthma as may worsen symptoms

(*Continued*)

Table 14.12 (*Contd.*)

Condition	Features	Management
Granulomatosis with polyangiitis (formerly Wegener's granulomatosis)	Granulomatous vasculitis Any organ may be involved; symptoms/signs relate to those affected, e.g. mouth ulcers; nasal ulceration with epistaxis/rhinitis; otitis media; cranial nerve lesions; lung symptoms and shadows on CXR; ↑ BP; eye signs (50%) Often long prodrome of 'limited granulomatosis with polyangiitis'—nasal stuffiness, headaches, hearing difficulties and nose bleeds	Refer to rheumatology/ general medicine for investigation ANCA helps diagnostically and in disease monitoring Treatment is with high-dose steroids, methotrexate, mycophenolate mofetil, and cyclophosphamide
Kawasaki's disease	Predominantly affects children <5y Cause unknown *Diagnosis:* diseases with similar presentations have been excluded and ≥5 of: ● Fever for ≥5d ● Bilateral conjunctivitis ● Polymorphous rash ● Changes in lips/mouth—red, dry, or cracked lips; strawberry tongue; diffuse redness of mucosa ● Changes in extremities: reddening of palms/soles; oedema of hands/feet; peeling of skin of hands, feet, and/or groin ● Cervical lymphadenopathy >15mm diameter (usually single and painful) ↑ suspicion if poor response to antipyretics	If suspected refer for urgent paediatric assessment Early treatment (<10d after onset) with IV immunoglobulin and aspirin ↓ incidence and severity of aneurysm formation as well as giving symptom relief Complications: Coronary arteritis with formation of aneurysms; accelerated atherosclerosis

Patient information and support

Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org
European Vasculitis Study Group 🖱 www.vasculitis.org
Kawasaki Support Group ☎ 024 7661 2178 🖱 www.kssg.org.uk
Vasculitis UK 🖱 www.vasculitis.org.uk

Tiredness and chronic fatigue syndrome

Tired all the time Fatigue is common. 1:400 sustained episodes of fatigue generate a GP consultation. GPs see 30 patients/y whose main complaint is fatigue and it may be a 2° symptom in many others. 2% of consultations result in 2° care referral. Almost any disease processes can cause tiredness—whether physical or psychological. Physical causes account for ~9% of cases; 75% have symptoms of emotional distress.

Assessment

- **Onset/duration** Short history/abrupt onset suggest post-viral or DM
- **Pattern of fatigue** On exertion relieved by rest suggests organic cause; worst in the morning and never goes suggests depression
- **Associated symptoms** e.g. breathlessness, weight ↓, or anorexia suggest underlying organic disease. Chronic pain may cause fatigue
- **Sleep patterns** Early morning wakening/unrefreshing sleep suggest depression; snoring, pauses of breathing in sleep, and daytime sleepiness suggest sleep apnoea (➔ p. 308)
- **Psychiatric history** Symptoms of depression, anxiety, and stress
- **Alcohol and medication** Including OTC and illicit drugs
- **Patient's worries** What does the patient think is wrong?
- **Examination** Usually normal

Common organic causes of fatigue in general practice

- Anaemia
- Infections (EBV, CMV, hepatitis)
- DM
- Hypo- or hyperthyroidism
- Perimenopausal
- Asthma
- Carcinomatosis
- Sleep apnoea

Investigation If sustained fatigue with no obvious cause, check:

- **Urine** Dipstick for protein, blood, and glucose
- **Blood** FBC (all children should have FBC/blood film checked on presentation with fatigue^{N}); ESR/CRP; U&E, Cr, and eGFR; LFTs and Ca^{2+}; TFTs; random blood glucose; anti-endomysial antibody test (to exclude coeliac disease); CK; consider Lyme disease titres

> In addition, check serum ferritin if the patient is a child/young person. Do not check ferritin in adults unless FBC suggests iron deficiency.

Use clinical judgement to decide on additional tests to exclude other diagnoses (e.g. serological testing if history suggestive of infection).

Management Treat organic causes. In most no physical cause is found—reassure. Explaining the relationship of psychological and emotional factors to fatigue can help patients deal with symptoms. If lasts >6–12wk and symptoms/signs of depression, consider a trial of antidepressants, e.g. sertraline 50mg od.

Chronic fatigue syndrome (CFS, ME) A debilitating and distressing condition. *Prevalence*: 0.2–2.6%; ♀:♂ ≈3:2. Cause is unknown though viral infections (≈10% after EBV), immunization, chemical toxins (e.g. organophosphates, chemotherapy drugs) have all been implicated.

❗ Fatigue must have been present for ≥4mo for adults and ≥3mo for children and young people for a diagnosis of CFS to be made.

Clinical features Unexplained persistent and/or recurrent fatigue of new/definite onset, not explained by other conditions and resulting in ↓ activity (often starting 1–2d after mental/physical exertion and lasting >24h) and ≥1 of:

- General malaise
- Dizziness/nausea
- Palpitations without cardiac dysfunction
- Cognitive dysfunction, e.g. impaired concentration/memory
- Tender cervical/axillary LNs without enlargement
- Physical/mental exertion makes symptoms worse
- Headaches of new type, pattern, or severity
- Multi-site muscle/joint pain without inflammation
- Sore throat
- Difficulty with sleeping

Additional symptoms Must not have pre-dated fatigue. Symptoms may fluctuate or change in nature over time. Include:

- Postural dizziness
- Vertigo
- Altered temperature sensation
- Paraesthesiae
- Sensitivity to light/sound
- Palpitations
- IBS
- Food intolerance
- Fibromyalgia
- Feelings of dyspnoea
- Mood swings
- Panic attacks
- Depression

❗ Infection/immunization, drugs, caffeine, alcohol, and stress cause setbacks.

⚠ *Red flag symptoms that suggest another diagnosis*

- Significant weight ↓
- Localizing/focal neurological signs
- Signs/symptoms of inflammatory arthritis or connective tissue disease
- Signs/symptoms of cardiorespiratory disease
- Sleep apnoea
- Clinically significant lymphadenopathy

Reconsider diagnosis if none of the following are present Cognitive difficulties; chronic pain; post-exertional fatigue/malaise; sleep disturbance.

Management Provide support and reassurance—explanation, information ± self-help groups. Avoid exacerbating factors, e.g. caffeine, alcohol. Advise graded exercise and regular, limited rest periods (e.g. 30min 4–5×/d). Treat symptoms, e.g. amitriptyline 10–50mg nocte to help sleep ± relieve headache/neuropathic pain; SSRI for depression. Refer adults if severe symptoms, or symptoms persist >6mo. Specialist treatments include CBT and rehabilitation programmes.

Prognosis Variable. 55% of adults have symptoms >6mo. Risk ↑ ×3 if history of anxiety/depression. Prognosis in children is better.

Children presenting with sustained fatigue of any duration with no obvious cause should *always* be referred for paediatric review

Further information

NICE (2007, expected update 2020) Diagnosis and management of CFS/ME in adults and children. 🖱 www.nice.org.uk/guidance/cg53

Information and support for patients

Action for ME ☎ 0117 927 9551 🖱 www.actionforme.org.uk
ME Association ☎ 0844 576 5326 🖱 www.meassociation.org.uk

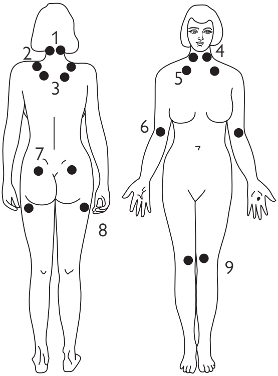
Miscellaneous conditions

Neuropathic arthritis *Charcot's disease* is a rapidly progressive degeneration in a joint which lacks position sense and protective pain sensation. Upper limb disease is usually associated with syringomyelia. Lower limb disease is usually associated with diabetic neuropathy or cauda equina lesions. The joint may be very deformed but is usually painless. Treat the underlying condition (e.g. DM). The joint cannot recover but refer to orthopaedics for advice on stabilization.

Fibromyalgia Painful, non-articular condition of unknown cause, predominantly involving muscles. Fibromyalgia is common and often results in significant disability. Peak age 40–50y; 90% female.

Diagnostic criteria

- History of widespread pain (defined as pain on both left and right sides, above and below the waist, together with axial skeletal pain, e.g. neck or back pain), *in combination with*
- Pain in ≥11 out of 18 tender sites (Figure 14.9) on digital palpation



1. Insertion of nuchal muscles into the occiput
2. Upper border of trapezius mid-portion
3. Muscle attachments to upper medial border of scapula
4. Anterior aspects of the C_5, C_7 intertransverse spaces
5. Second rib space ~3cm lateral to the sternal border
6. Muscle attachments to the lateral epicondyle at the elbow
7. Upper outer quadrant of gluteal muscles
8. Muscle attachments just posterior to the greater trochanter
9. Medial fat pad of the knee just proximal to the joint line

Figure 14.9 Tender point sites for diagnosis of fibromyalgia

Other clinical features

- Clinical findings are unremarkable
- Pain is worsened by stress, cold, and activity and associated with generalized morning stiffness; analgesics, NSAIDs, and local physical treatments are ineffective and may worsen symptoms
- Sleep patterns are poor—patients tend to wake exhausted and complain of poor concentration; anxiety and depression scores are high
- Associated symptoms include unexplained headache, paraesthesiae of hands/feet, urinary frequency, and abdominal symptoms

Investigation Exclude other causes of pain and fatigue (e.g. hypothyroidism, SLE, Sjögren's, psoriatic arthritis, inflammatory myopathy, hyperparathyroidism, osteomalacia)—check FBC, ESR, TFTs, U&E, eGFR, Ca^{2+}, CK, PO_4, ANA, RF, and immunoglobulins.

Management Multidisciplinary approach is helpful.

- Be supportive—reassurance that there is no serious pathology, explanation, and information are vital
- Low-dose amitriptyline 25–75mg nocte may help with sleep/pain. SSRI, e.g. sertraline 25–50mg od may help anxiety, depression, and sleep—stop if no improvement after a month's trial
- Graded exercise regimes improve pain, lethargy, mood, and malaise
- Counselling/learning coping strategies/CBT can be beneficial
- 💣 Some patients benefit from injection of hyperalgesic trigger points with steroid or acupuncture to trigger points

Hypermobility Children/young adults with lax joints; <50% are symptomatic. Those that have symptoms present with recurrent joint pains—mainly affecting the knees. Other symptoms include joint effusion, dislocation, ligamentous injuries, low back pain, and premature OA. The condition is benign, and joints become stiffer with age. Treatment, when needed, is with physiotherapy. Rarely associated with congenital disorders, e.g. Ehlers–Danlos syndrome.

Tietze syndrome Idiopathic costochondritis. Pain is enhanced by motion, coughing, or sneezing. The 2nd rib is most commonly affected. *Examination*: marked localized tenderness. *Differential diagnosis*: muscular sprain; rarely inflammatory chest wall enthesitis/osteitis 2° to spondyloarthropathy.

Management Explanation and reassurance that nothing serious is happening; simple OTC analgesia, e.g. ibuprofen 400mg tds. If pain persists local steroid or bupivacaine injections can be helpful. If not settling, consider referral to rheumatology.

Further information

European League Against Rheumatism (EULAR) (2016) Revised recommendations for the management of fibromyalgia. 🖱 http://ard.bmj.com/content/early/2016/07/04/annrheumdis-2016-209724

Patient information and support

Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org
Fibromyalgia Association UK ☎ 0300 999 3333 🖱 wwww.fmauk.org
Hypermobility Syndrome Association (HMSA) ☎ 0333 011 6388 🖱 http://hypermobility.org/

Chapter 15

Neurology

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Reflexes and muscle power

Reflexes Reflexes are automatic responses. The reflex arc goes from the stimulus via a sensory nerve to the spinal cord and then back along a motor nerve to cause muscle contraction, without brain involvement.

Key reflexes Table 15.2. Record whether absent, present with reinforcement, normal, or brisk ± clonus.

Absent or ↓ reflex Implies a breach in the reflex arc at:

- Sensory nerve or root, e.g. neuropathy, spondylosis
- Anterior horn cell, e.g. MND, polio
- Motor nerve or root, e.g. neuropathy, spondylosis
- Nerve endings, e.g. myasthenia gravis, or
- Muscle, e.g. myopathy

↑ reflex Implies lack of higher control—an upper motor neuron (UMN) lesion, e.g. post stroke

Clonus Rhythmic involuntary muscle contraction due to abrupt tendon stretching, e.g. by dorsiflexing the ankle—associated with an UMN lesion.

Reinforcement Method of accentuating reflexes. Use if a reflex seems absent. Ask the patient to clench their teeth (to reinforce upper limb reflexes) or clench their hands and pull in opposite directions (to accentuate lower limb reflexes). This effect only lasts ~1sec, so ask the patient to perform the manoeuvre simultaneously with the tap from the tendon hammer.

Testing for muscle power Table 15.1

Table 15.1 Quick screening test for muscle power

Joint	Movement	Nerve roots	Joint	Movement	Nerve roots
Shoulder	Abduction	C5, C6	*Hip*	Flexion	L1–3
	Adduction	C6–8		Extension	L4, L5, S1
Elbow	Flexion	C5, C6	*Knee*	Flexion	L5, S1
	Extension	C7, C8		Extension	L3, L4
Wrist	Flexion	C7, C8	*Ankle*	Dorsiflexion	L4, L5
	Extension	C6, C7		Plantarflexion	S1, S2
Fingers	Flexion	C8	*Toes*	Extensors	L5, S1
	Extension	C7		Flexors	S2
	Abduction	T1			

❗ Test proximal muscle power by asking the patient to sit from lying, pull you towards him/herself or rise from squatting.

Table 15.2 Key reflexes and nerve roots involved

Reflex	Test	Expected result	Nerve roots
Jaw	Ask the patient to let his mouth open slightly. Place a finger on the chin and tap the finger with a tendon hammer	Contraction of masseters and closure of mouth	Vth cranial nerve
Gag	Touch the back of the patient's pharynx on each side with a spatula. If absent, ask the patient whether he can feel the spatula—if he can, then Xth nerve palsy	Contraction of the soft palate	IXth/Xth cranial nerve
Biceps	Tap a finger placed on the biceps tendon by letting the tendon hammer fall on it	Contraction of the biceps + elbow flexion	C5, C6
Supinator	Tap the lower end of the radius just above the wrist with the tendon hammer	Contraction of brachioradialis + elbow flexion	C5, C6
Triceps	Support elbow in flexion with one hand. Tap the triceps tendon with a tendon hammer held in the other hand	Contraction of triceps + elbow extension	C7, C8
Knee	Support the knees so relaxed and slightly bent. Let the tendon hammer fall onto the infrapatellar tendon	Contraction of quadriceps + extension of knee	L3, L4
Ankle	Externally rotate the thigh and flex the knee. Let the tendon hammer fall onto the Achilles tendon	Contraction of gastrocnemius + plantar flexion of the ankle	S1
Abdominal	Lightly stroke the abdominal wall diagonally towards the umbilicus in each of the four abdominal quadrants	Abdominal wall contractions. When absent can be normal or indicate UMN or LMN lesion	T7–T12
Cremaster	♂ patients only. Pre-warn the patient. Stroke the superior and medial aspect of the thigh in a downwards direction	Contraction of cremasteric muscle → raising of scrotum and testis on the side stroked. Absent in UMN and LMN lesions	L1
Anal	Scratch the perianal skin	Reflex contraction of the external sphincter. Absent in UMN and LMN lesions	S4, S5
Plantar	Pre-warn the patient. Run a blunt object up the lateral side of the sole of the foot, curving medially before the MTP joints	Flexion of big toe (if >1y old). Extension implies UMN lesion.	S1

Cranial nerve lesions

Cranial nerves may be disrupted at any point from the nerve nucleus in the brainstem to the point of innervation. Table 15.3 lists clinical tests for each nerve; Figure 15.1 maps cutaneous innervation of the head/neck. Think systematically about the level of the lesion. *Potential sites*:

- Muscle
- Neuromuscular junction
- Along the course of the nerve outside the brainstem
- Within the brainstem

Any cranial nerve may be affected by DM, MS, tumours, sarcoid, vasculitis, or syphilis and >1 nerve may be affected by a lesion. Refer according to cause (usually to ENT, ophthalmology, or neurology).

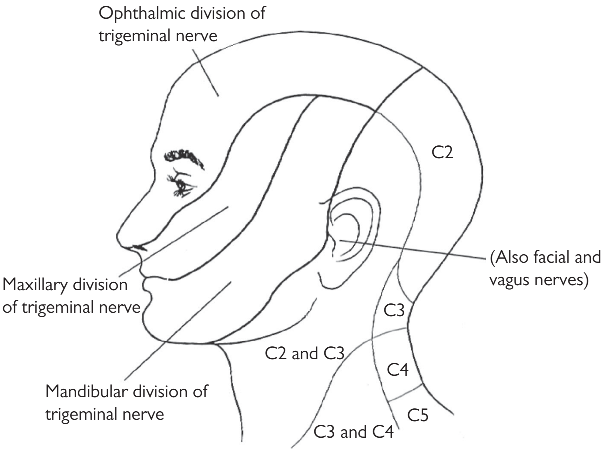
Table 15.3 Cranial nerve lesions and their causes

Nerve	Clinical test	Causes
I Olfactory	*Smell*—test each nostril for the ability to differentiate different smells	Trauma, frontal lobe tumour, meningitis
II Optic	*Acuity*—Snellen chart *Visual fields*—compare with your own visual fields by standing directly in front of the patient with your head at the same level as theirs	*Monocular blindness*—lesion in one eye or optic nerve (e.g. MS, giant cell arteritis)
	Pupils—size, shape, reaction to light, and accommodation *Ophthalmoscopy*—darken room, view optic disc (?pale, swollen), follow each vessel outwards to view each quadrant, track outwards to check lens and cornea	*Bitemporal hemianopia*—optic chiasm compression, e.g. pituitary adenoma, craniopharyngioma, internal carotid artery aneurysm *Homonymous hemianopia*—affects half the visual field on the side opposite the lesion. Caused by lesion beyond the optic chiasm, e.g. stroke, abscess, tumour
III	Ptosis, large pupil, eye looks down- and outwards ❗ Diplopia from a IIIrd nerve lesion may cause nystagmus	DM, giant cell arteritis, syphilis, posterior communicating artery aneurysm, idiopathic If pupil normal size, results from DM or other vascular cause
IV	Diplopia on looking down and in; may compensate by tilting head	Rare in isolation. May occur as a result of trauma to the orbit
V Trigeminal	*Motor*—open mouth. Jaw deviates to the side of the lesion *Sensory*—corneal reflex lost first. Check all 3 divisions	*Sensory*—trigeminal neuralgia (➔ p. 531); herpes zoster, nasopharyngeal carcinoma *Motor*—bulbar palsy (➔ p. 519), acoustic neuroma
VI	Horizontal diplopia on looking outwards	MS, pontine CVA, ↑ ICP

(*Continued*)

Table 15.3 (*Contd.*)

Nerve	Clinical test	Causes
VII Facial	Causes facial weakness and droop Ask to raise eyebrows, show teeth, puff out cheeks. • *LMN lesion*: all one side of face affected • *UMN lesion*: lower two-thirds face affected only	*LMN*—Bell's palsy, polio, otitis media, skull fracture, cerebellopontine angle tumour, parotid tumour, herpes zoster (Ramsay Hunt syndrome ➔ p. 512) *UMN*—stroke, tumour
VIII Vestibulo-auditory	*Auditory*—ask to repeat a number whispered in 1 ear while you block the other *Vestibular*—ask about balance, check for nystagmus (➔ p. 928)—ask patient to fix on finger 0.75 m away—check gaze upwards, downwards, lateral (both directions), keeping finger <30° from midline	Noise, Paget's disease, Ménière's disease (➔ p. 929), herpes zoster, acoustic neuroma, brainstem CVA, drugs (e.g. furosemide)
IX, X	Gag reflex, palate moves → normal side on saying 'Aah'	Trauma, brainstem lesions, neck tumours
XI	*Trapezii*—shrug shoulders against resistance *Sternomastoid*—turn head to right/left against resistance	**Rare.** Polio, syringomyelia, tumours near jugular foramen, stroke, bulbar palsy (➔ p. 519), polio, trauma, TB
XII	Tongue deviates to the side of the lesion	Trauma, brainstem lesions, neck tumours



Ophthalmic division of trigeminal nerve
C2
(Also facial and vagus nerves)
Maxillary division of trigeminal nerve
C3
C2 and C3
C4
Mandibular division of trigeminal nerve
C5
C3 and C4

Figure 15.1 Cutaneous innervation of the head and neck

Neuropathy

Dermatomes and peripheral nerve distribution Figure 15.2 (➔ p. 514) and Figure 15.3 (➔ p. 515)

Mononeuropathy Lesions of individual peripheral (including cranial) nerves. *Causes*: trauma, compression, DM, leprosy. If >1 peripheral nerve is involved, the term *mononeuritis multiplex* is used. *Causes*: DM, sarcoid, cancer, PAN, amyloid, leprosy.

Common mononeuropathies Table 15.4

Bell's palsy Facial palsy without other signs. Unknown cause—possibly viral. *Peak age*: 10–40y. ♂ = ♀. *Lifetime incidence*: ~1 in 65. Affects left and right side of the face equally often. Usually sudden onset—may be preceded by pain around the ear. *Other possible symptoms*: facial numbness; ↓ noise tolerance; disturbed taste on the anterior part of the tongue.

Management ~70% recover completely; 13% have insignificant sequelae; the remainder have permanent deficit. 85% improve in <3wk—reassure. Give prednisolone (25mg bd for 10d) if <72h after onset of symptoms[R]. Protect eye—tape lid shut and pad at night; glasses in the day ± artificial tears if drying. *Refer*:

- If recovery is not starting after 3wk
- For tarsorrhaphy if complete or longstanding palsy
- If unacceptable cosmetic result—may benefit from plastic surgery

Ramsay Hunt syndrome (herpes zoster oticus) Severe pain in the ear precedes facial nerve palsy. Zoster vesicles appear around the ear, in the external ear canal, on the soft palate, and in the tonsillar fossa. Often accompanied by deafness ± vertigo which are slow to resolve and may result in permanent deficit. Pain usually abates after 48h but post-herpetic neuralgia can be a problem. If detected <24h after the rash appears, treatment with antivirals (e.g. aciclovir 800mg 5×/d for 1wk) may be effective.

Morton's metatarsalgia ➔ p. 472

Autonomic neuropathy Postural hypotension (dizziness or syncope on standing, or after exercise/large meal), impotence, inability to sweat, vomiting and dysphagia, diarrhoea or constipation, urinary retention or incontinence, Horner's syndrome (➔ p. 270). Check BP lying and standing—a postural drop of ≥30/15mmHg is abnormal. *Causes*:

- **Primary autonomic failure** No known cause. Occurs alone or as part of multisystem atrophy. Typically middle-aged/elderly men. Onset is insidious. Survival—rarely >10y after diagnosis
- **Ageing** 25% >74y have postural hypotension. Review medication, discourage prolonged bed rest. Often associated with disordered thermoregulation making elderly people prone to hypothermia. Exclude other disorders (e.g. DM, multisystem atrophy, drugs) before putting down to ageing alone
- **Drugs** Common culprits—antihypertensives (e.g. thiazides), diuretics (over-diuresis), levodopa, TCAs, phenothiazines, benzodiazepines
- **Polyneuropathies** May be part of more general polyneuropathy, e.g. DM, Guillain–Barré syndrome, or alcoholic/nutritional neuropathy

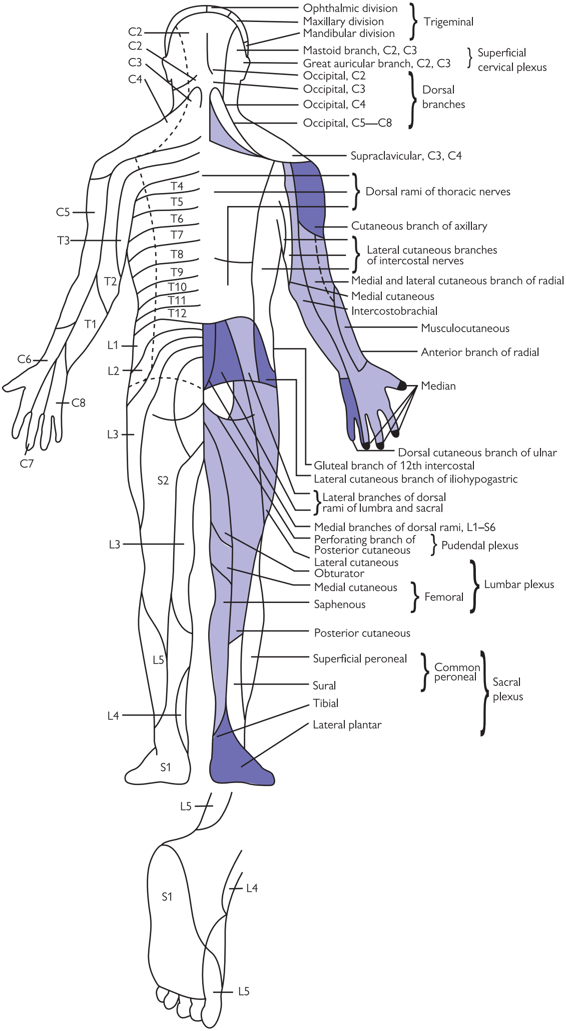
- Other causes Craniopharyngioma, vascular lesions, spinal cord lesions, tabes dorsalis, Chagas' disease, HIV, familial dysautonomia

Management Treat any underlying cause. Advise patients to stand slowly, raise the head of the bed at night, eat little and often and ↓ carbohydrate and alcohol intake. Fludrocortisone (0.1mg/d, increasing prn) may help those most severely affected. Refer if diagnosis is unclear or simple measures are ineffective.

Polyneuropathy ➔ p. 516

Table 15.4 Common mononeuropathies

Nerve involved	Nerve roots	Presentation	Common causes
Median	C5–T1	Loss of sensation over lateral 3½ fingers and palm Wasting of the thenar eminence Inability to flex the terminal phalanx of the thumb implies involvement of the anterior interosseous branch	Trauma (especially wrist lacerations), carpal tunnel syndrome (➔ p. 460)
Ulnar	C7–T1	Weakness and wasting of interossei muscles (weakness of abduction of fingers) and claw hand deformity Wasting of hypothenar eminence, sensory loss over medial 1½ fingers and ulnar side of the hand Flexion of 4th and 5th fingers is weak if proximal lesion	Trauma or compression at the elbow (➔ p. 456), trauma at the wrist
Radial	C5–T1	Sensory loss is variable but always includes the dorsal aspect of the root of the thumb Wrist drop and weak extension of thumb and fingers	Compression against the humerus, trauma
Sciatic	L4–S2	Weakness of hamstrings and all muscles below the knee (foot drop) Loss of sensation below the knee laterally	Back injury, pelvic tumour
Common peroneal	L4–S2	Inability to dorsiflex the foot (foot drop), evert the foot, or extend the toes. Sensory loss over dorsum of the foot	Trauma
Tibial	S1–S3	Inability to stand on tiptoe, invert the foot or flex toes Sensory loss over sole	Trauma or entrapment


Ophthalmic division
Maxillary division
Mandibular division
Trigeminal
Mastoid branch, C2, C3
Great auricular branch, C2, C3
Superficial cervical plexus
Occipital, C2
Occipital, C3
Occipital, C4
Occipital, C5—C8
Dorsal branches
Supraclavicular, C3, C4
Dorsal rami of thoracic nerves
Cutaneous branch of axillary
Lateral cutaneous branches of intercostal nerves
Medial and lateral cutaneous branch of radial
Medial cutaneous
Intercostobrachial
Musculocutaneous
Anterior branch of radial
Median
Dorsal cutaneous branch of ulnar
Gluteal branch of 12th intercostal
Lateral cutaneous branch of iliohypogastric
Lateral branches of dorsal rami of lumbra and sacral
Medial branches of dorsal rami, L1–S6
Perforating branch of Posterior cutaneous
Pudendal plexus
Lateral cutaneous
Obturator
Medial cutaneous
Saphenous
Femoral
Lumbar plexus
Posterior cutaneous
Superficial peroneal
Sural
Common peroneal
Tibial
Lateral plantar
Sacral plexus
C2
C2
C3
C4
C5
T3
T2
T1
C6
C8
C7
T4
T5
T6
T7
T8
T9
T10
T11
T12
L1
L2
L3
S2
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L5
L4
S1
L5
S1
L4
L5

Figure 15.2 Dermatomes and peripheral nerve distribution

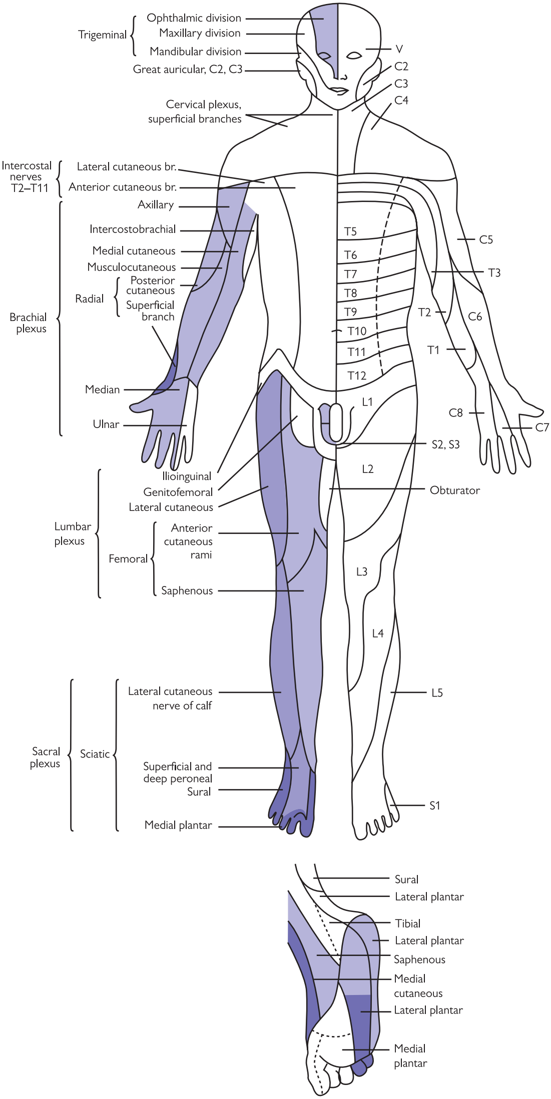
Trigeminal
Ophthalmic division
Maxillary division
Mandibular division
Great auricular, C2, C3
Cervical plexus, superficial branches
V
C2
C3
C4
Intercostal nerves T2–T11
Lateral cutaneous br.
Anterior cutaneous br.
Brachial plexus
Axillary
Intercostobrachial
Medial cutaneous
Musculocutaneous
Radial
Posterior cutaneous
Superficial branch
Median
Ulnar
T5
T6
T7
T8
T9
T10
T11
T12
C5
T3
T2
C6
T1
C8
C7
L1
S2, S3
L2
Obturator
Lumbar plexus
Ilioinguinal
Genitofemoral
Lateral cutaneous
Femoral
Anterior cutaneous rami
Saphenous
L3
L4
L5
Sacral plexus
Sciatic
Lateral cutaneous nerve of calf
Superficial and deep peroneal
Sural
Medial plantar
S1
Sural
Lateral plantar
Tibial
Lateral plantar
Saphenous
Medial cutaneous
Lateral plantar
Medial plantar

Figure 15.3 Dermatomes and peripheral nerve distribution

Polyneuropathy

Generalized disorder of peripheral nerves, including cranial and autonomic nerves. Distribution is bilateral, symmetrical, and widespread.

Sensory neuropathy Presents as numbness, tingling, or burning sensation often affecting the extremities first (glove and stocking distribution) or causing clumsiness handling fine objects (e.g. needle).

Motor neuropathy Presents as progressive weakness or clumsiness of hands, stumbling/falls on walking, respiratory difficulty (can progress rapidly). *Examination*: wasting and weakness most marked distally; reflexes are ↓ or absent.

Causes Table 15.5

Initial investigations Exclude common causes—check blood glucose, FBC, ESR, U&E, Cr and eGFR, LFTs, TFTs, plasma vitamin B_{12}, autoimmune profile, syphilis serology.

Management Treat cause if possible. Involve physiotherapists and OT. If sensory neuropathy care of the feet is important to minimize trauma and consequent disability. Refer if a cause is not found.

⚠ If rapid deterioration, admit as acute medical emergency as ventilation may be needed.

Specific polyneuropathies

Charcot–Marie–Tooth syndrome (peroneal muscular atrophy) Presents at puberty or in early adult life and begins with foot drop and weak legs. The peroneal muscles are the first to atrophy. The disease spreads to the hands then arms. Sensation and reflexes are also ↓. Unknown cause. Once diagnosis is confirmed, treatment is supportive.

Guillain–Barré polyneuritis Develops within a few weeks of surgery, 'flu vaccination, or infection (URTI, flu, varicella zoster, HSV, CMV, EBV, *Campylobacter, Mycoplasma*, Zika). In 40% no precipitating event is found.

Presents with ascending motor neuropathy which may advance fast. Proximal muscles are more affected than distal muscles. Trunk, respiratory muscles, and cranial nerves are commonly affected.

If suspected admit immediately to hospital as an emergency. Ventilation on ITU is frequently required; 85% make a complete or near complete recovery; 10% are unable to walk alone at 1y; mortality is 10%.

Polio ➔ p. 551

Refsum's syndrome Rare autosomal recessive disorder which presents in the 2nd decade or later with sensorimotor polyneuropathy, ataxia, visual and/or hearing problems. Treatment involves dietary restriction (avoidance of chlorophyll-containing foods) and plasmapheresis.

Table 15.5 Causes of polyneuropathy

Inflammatory	Guillain–Barré syndrome (mostly motor) Chronic inflammatory demyelinating polyneuropathy (CDP) Sarcoidosis
Metabolic	DM (mainly sensory) Renal failure (mainly sensory) Hypothyroidism Hypoglycaemia Mitochondrial disorders
Vasculitis	Polyarteritis nodosa Rheumatoid arthritis Granulomatosis with polyangiitis
Malignancy	Paraneoplastic syndromes (especially small cell lung cancer) Polycythaemia rubra vera
Infection	HIV Syphilis Lyme disease Leprosy (mainly sensory)
Vitamin deficiency	Lack of vitamins B_1, B_6, B_{12} (e.g. alcoholic)
Inherited	Refsum's syndrome Charcot–Marie–Tooth syndrome (mostly motor) Porphyria
Toxins	Lead (mostly motor) Arsenic
Drugs	Alcohol Cisplatin Isoniazid Vincristine Nitrofurantoin *Less frequently*: metronidazole, phenytoin
Others	Paraproteinaemias, e.g. multiple myeloma, amyloidosis

Speech problems

Hoarseness ➲ p. 914

Stammer Disorder of rhythm and fluency of speech in which syllables, words, or phrases are repeated. ♂:♀ ≈4:1. *Cause*: unknown. Can result in stress and embarrassment.

- Younger children Often short-lived; usually resolves spontaneously
- Older children/adults Refer to speech therapy

Dysarthria Difficulty with articulation due to incoordination or weakness of the musculature of speech. Language is normal. Ask to repeat 'baby hippopotamus' or 'British constitution'. Treat the cause if possible, otherwise support with speech therapy and aids to communication. *Assessment and causes*: Table 15.6.

Dysphasia Impairment of language due to brain damage to the dominant hemisphere. The left hemisphere is dominant for 99% of right-handed people and 60% of left handers. In most cases due to stroke or brain tumour. Rarely due to head injury or dementia. *Classification*: Table 15.7. Mixed pictures are common. *Treatment*:

- Speech therapy may or may not be helpful
- Support, e.g. dysphasia groups
- Aids to communication, e.g. computers, picture boards

Myasthenia gravis Autoimmune disease. Antibodies to the acetylcholine receptor cause a deficit of receptors at the neuromuscular junction → muscle weakness. Antibodies are detectable in 90%. ♀:♂ ≈2:1. Associated with thymic tumours and other autoimmune disease, e.g. RA, SLE, hyperthyroidism. Generally follows a relapsing or slowly progressive course. If thymoma present, 5y survival ≈30%.

Presentation Young adults with easy fatigability of muscles. Commonly affected muscles are the:

- Orbital muscles causing ptosis and diplopia *and*
- Bulbar muscles causing slurring of speech—ask to count to 50

Weakness is exacerbated by pregnancy, infection, drugs (e.g. β-blockers, opioids, tetracycline, quinine), climate change, emotion, and exercise.

Management If suspected, refer for confirmation by a neurologist and specialist treatment. *Treated with*:

- Anticholinesterase, e.g. pyridostigmine
- Immunosuppression with prednisolone, methotrexate, or azathioprine
- Thymectomy → remission in 30% and benefit in another 40%
- Plasmapheresis

Lambert–Eaton syndrome (or myasthenic syndrome) Occurs in association with small cell carcinoma of the lung or rarely autoimmune disease. Differs from myasthenia gravis by the tendency to hyporeflexia. Autonomic involvement is common. Proximal limb muscles/trunk are most commonly involved. Specialist treatment is essential.

Patient support

Myaware 🖱 www.myaware.org

Table 15.6 Causes of dysarthria

Cause	Characteristics
Cerebellar disease	Slurring of speech as if drunk Speech is irregular in volume and scanning in quality
Extrapyramidal disease e.g. Parkinson's disease	Soft, indistinct, and monotonous speech
Pseudo-bulbar palsy e.g. stroke (bilateral), MS, MND	Alteration of speech—typically nasal speech sounding like Donald Duck Difficulty swallowing or chewing Tongue is spastic and jaw jerk ↑
Bulbar palsy e.g. MND, Guillain–Barré, alcoholic brainstem myelinolysis, 1° or 2° brainstem tumours, syringobulbia, polio, hyponatraemia	Speech—quiet, hoarse, or nasal Loss of function of the tongue, muscles of chewing/swallowing ± facial muscles Flaccid, fasciculating tongue Jaw jerk normal or absent
Palate paralysis	Nasal speech Asymmetric or absent gag reflex
Myasthenia gravis	Slurring of speech when fatigued

Table 15.7 Assessment and classification of dysphasia

Assessment:

Is speech fluent, grammatical, meaningful, and apt? If yes, dysphasia is unlikely.

Comprehension: can the patient follow 1, 2, or multiple step commands?

Repetition: can the patient repeat a phrase after you?

Naming: can the patient name common and uncommon items?

Reading and writing? Usually affected too. If not, question the diagnosis of dysphasia.

Characteristics of dysphasia	Broca's (expressive)	Wernicke's (receptive)	Conduction	Transcortical
Fluent?	✗	✓	✓	✓ or ✗
Repetition normal?	✗	✗	✗	✓
Understanding impaired?	✗	✓	✗	✓ or ✗

Walking problems

Walking difficulty ('off legs') Common symptom in the elderly. *Causes*:

- Musculoskeletal Osteoarthritis or RA, osteoporotic fractures, fractured neck of femur, osteomalacia, Paget's disease, polymyalgia rheumatica
- Psychological Depression, bereavement, fear of falling
- Neurological Stroke, Parkinson's disease, peripheral neuropathy
- Spinal cord compression
- Systemic Pneumonia, UTI, anaemia, hypothyroidism, renal failure, infection, hypothermia

Management Treat according to cause. Refer if inadequate support at home, cause warrants admission, or no cause is found.

Abnormal gait Gait means manner of walking. Abnormal gait can give clues to the underlying problem.

Abnormal movements Normal gait is interrupted by abnormal movements, e.g. choreiform movements, athetoid movements, or hemiballismus. May indicate underlying neurological problem, e.g. cerebral palsy, Huntington's chorea.

Antalgic gait Gait adjusts to try to minimize pain in a joint—usually OA hip. The patient leans towards the affected side and takes a rapid step on that side followed by a slower step on the contralateral side—check Trendelenburg's sign (➔ p. 462).

Drunken gait Apart from alcohol, the other major cause of drunken gait is a cerebellar lesion. *Features*:

- Wide-based gait or reeling gait on a narrow base
- Feet are often raised too high and placed over carefully with the patient looking ahead
- If a cerebellar lesion, the patient falls to the side of the lesion

Foot drop Patients trip frequently or walk with a high-stepping gait. On examination, patients are unable to walk on their heels and cannot dorsiflex their foot. Check ankle jerk. *Causes*:

- Common peroneal palsy, e.g. due to trauma—normal ankle jerk
- Sciatica—ankle jerk absent
- L4, L5 root lesion—ankle jerk may be absent
- Peripheral motor neuropathy, e.g. alcoholic—ankle jerk weak or absent
- Distal myopathy—ankle jerk weak or absent
- MND—↑ ankle jerk

Hemiplegic gait Style of walking seen in patients with UMN lesions. *Features*:

- Arm adducted and internally rotated, elbow flexed and pronated ± finger flexion
- Foot is plantar flexed and the leg swings in a lateral arc

Frontal lesions Marked unsteadiness—the feet appear stuck to the floor causing a wide-based, shuffling gait.

Parkinsonian gait Seen in patients with Parkinson's disease and other causes of Parkinsonism. *Features*:

- **Shuffling gait** Short steps, with the feet barely leaving the ground, producing an audible shuffling noise. May trip over small obstacles
- **Turning 'en bloc'** Keeping the neck and trunk rigid, and requiring multiple small steps to accomplish a turn
- **Gait freezing** Inability to move feet. May worsen in tight, cluttered spaces or when attempting to initiate gait
- **Festinant gait** Flexed posture as if hurrying to keep up with feet
- **Lack of normal arm swing**

Scissor gait As the name implies, the patient walks as if their legs were like a pair of scissors. Associated with spastic paraplegia.

- Both legs are held rigid with plantar flexion of the ankle, extension of the knee, and adduction/internal rotation of the hips
- The patient walks on tiptoe and the knees rub together/cross during the walking cycle
- Often accompanied by complex movements of the upper limbs to assist the walking movements

Sensory ataxic gait Loss of proprioception due to peripheral neuropathy or spinal cord disease (e.g. cervical spondylosis, MS, syphilis, combined degeneration of the cord) results in an ataxic gait similar to that seen with cerebellar disease. Check Romberg's test. *Features*:

- Broad-based gait with a tendency to stamp feet down clumsily
- Patient tends to look at feet throughout the walking cycle
- Romberg's sign +ve

Waddling gait Typically seen in patients with proximal myopathy, e.g. due to muscular dystrophy. *Other causes*: pregnancy, congenital dislocation of the hip. *Features*:

- Broad-based gait. The pelvis drops to the side of the leg being raised
- The patient moves their body and hips to accommodate this resulting in a duck-like waddle in the swing phase
- Commonly accompanied by ↑ forward curvature of the lower spine

Examining gait Note abnormalities and any aids/assistance required.

- Make sure that you can see the legs well
- Ask the patient to stand up from a chair without support. If able to do that, repeat with feet together and/or with eyes closed
- Ask the patient to stand still with feet together. If able to do that, ask the patient to close their eyes and see what happens (*Romberg's sign*)
- Ask the patient to walk normally for ~5m, turn round, and walk back
- Ask the patient to walk heel-to-toe (testing for cerebellar disease)
- Ask the patient to stand with the feet together:
  - With eyes open—testing cerebellar and posterior column function
  - With eyes closed—testing posterior column function
  - On toes alone—impossible with S1 lesions
  - On heels alone—impossible if L4/L5 lesion

Other movement problems

Abnormal gait ➔ p. 520

Cramp Painful muscle spasm. Common—especially at night and after exercise. Rarely associated with disease—salt depletion, muscle ischaemia, myopathy. Forearm cramps suggest MND. Night cramps may respond to quinine bisulfate 300mg nocte 2× weekly for <3mo.

Dystonia Prolonged muscle contraction producing abnormal postures or repetitive movements.

- Spasmodic torticollis Head is pulled to one side and held there by a contracting sternomastoid muscle. Treat with massage, heat, analgesia, ± physiotherapy
- Blepharospasm Involuntary contraction of the orbicularis oculi. If troublesome, consider short-term treatment with diazepam (but be careful to avoid dependence) or refer for treatment with botulinum toxin
- Writer's cramp Spasm of the hand and forearm muscles on writing
- Generalized dystonia Primary generalized dystonia is usually genetic. Specialist treatment from a neurologist is essential. First-line drug treatment is with levodopa. If that is ineffective, an anticholinergic drug (e.g. trihexyphenidyl) can be helpful in controlling muscle spasms and tremor. Other second-line treatments include clonazepam, tetrabenazine, baclofen, botulinum toxin injections. Deep brain stimulation may also be helpful
- Secondary dystonia Symptoms of dystonia that result from drugs or other medical conditions. Includes drug-induced dystonia (acute dystonia or tardive dyskinesia); dystonia associated with cerebral palsy; dystonia associated with Parkinson's disease; dystonia associated with other brain injury or disease; and dystonia associated with metabolic conditions (e.g. Wilson's disease)

Tardive dyskinesia Involuntary chewing and grimacing movements resulting from long-term neuroleptic treatment (e.g. chlorpromazine); metoclopramide and prochlorperazine are also possible causes. Withdraw the drug implicated—if no improvement after 3–6mo consider tetrabenazine 25–50mg tds po.

Patient information and support

Dystonia Society ☎ 020 7793 3650 🖱 www.dystonia.org.uk

Myoclonus Sudden involuntary focal or general jerks. May be normal especially if occurs when falling asleep. *Other causes*:

- Neurodegenerative disease (e.g. CJD)
- Myoclonic epilepsy
- Benign essential myoclonus (generalized myoclonus beginning in childhood as muscle twitches, may be inherited as autosomal dominant)
- Asterixis (metabolic flap—e.g. liver failure, uraemia)

If needed, treat with sodium valproate or clonazepam—take specialist advice.

Dyspraxia Impairment of performance of complex movements despite preservation of ability to perform their individual components. Test by asking the patient to perform everyday tasks (e.g. ask to dress/undress), copy complex hand movements, and do familiar sequences of movements (e.g. 'head, shoulders, knees, and toes').

Childhood (developmental coordination disorder) ➔ p. 894

Adults Most common causes are stroke or space-occupying lesion. Involve rehabilitation services and OT.

- **Dressing dyspraxia** Unable to dress correctly
- **Constructional dyspraxia** Difficulty in assembling objects or drawing (ask to draw 5-pointed star)
- **Gait dyspraxia** Gait disorder although the lower limbs function normally—more common among the elderly

Tremor

- **Resting tremor** Present at rest but abolished on voluntary movement. Most common cause—PD when tremor is rhythmic
- **Intention tremor** Irregular large amplitude tremor worse on movement, e.g. reaching for something. Typical of cerebellar disease
- **Tremors on movement** Thyrotoxicosis, anxiety, benign essential tremor (inherited), and drugs (e.g. β-agonists) cause a fine tremor abolished at rest. Alcohol and β-blockers may help

Asterixis Intermittent lapses of an assumed posture. May involve arms, neck, tongue, jaw, and eyelids. Usually bilateral, absent at rest, and asynchronous on each side. *Causes*: liver failure (flapping tremor), heart failure, respiratory failure, renal failure, hypoglycaemia, barbiturates.

Athetosis Slow, confluent, often rhythmic, purposeless movements of hands, tongue, fingers, or face. *Causes*: cerebral palsy, kernicterus.

Chorea Non-rhythmic, jerky, purposeless movements (especially hands) with voluntary movements possible in between. *Most common causes*: cerebral palsy, Huntington's chorea, Sydenham's chorea.

Ballismus/hemiballismus Large-amplitude, involuntary flinging movements of limbs. May occur after stroke, in Huntington's disease, or with high doses of levodopa for PD.

Tics Brief, repeated, and stereotyped movements which are able to be suppressed voluntarily for a while. Common in children and usually resolve spontaneously. Consider clonazepam or clonidine if tics are severe.

Gilles de la Tourette syndrome ➔ p. 889

Dizziness and giddiness Distinguish between true vertigo (the illusion of rotatory movement—the room spinning) and a feeling of unsteadiness or lightheadedness:

- **Vertigo** ➔ p. 928
- **Imbalance** Implies difficulty in walking straight, e.g. from disease of peripheral nerves, posterior columns, or cerebellum
- **Faintness** The feeling of being about to pass out. Associated with some seizure disorders and a variety of non-neurological conditions (e.g. postural hypotension, vasovagal syncope; hyperventilation; hypoglycaemia; arrhythmias). Sometimes >1 element coexists

Transient loss of consciousness

Transient loss of consciousness in the form of blackouts, faints, and funny turns is a common presentation to general practice.

Funny turns in small children ➲ p. 874

History A good history from the patient and ideally from a witness too is essential to establish the correct diagnosis. *Ask*:

- Did the patient lose consciousness? Quite frequently patients describe episodes of dizziness or unsteadiness/falling as 'funny turns'
- Does the patient remember the whole episode? If not, which bits are missing and how long are the gaps?
- What happened before, during, and after the event? Duration of event?
- When and where? Posture at the time of the event (standing, sitting, lying). Did it start during sleep? Were there any precipitating events?
- Were there any warning signs (e.g. aura, feeling going to faint, sweating, feeling hot, visual disturbance, etc.)?
- Did the patient jerk his/her limbs? If so, was the jerking generalized or restricted to one area of the body?
- What did the patient look like? Eyes open/shut; colour?
- Did anything else happen (e.g. tongue biting; incontinence)?
- What happened after the attack? Was the patient conscious straight away? Was there disorientation/confusion, drowsiness, weakness down one side?

Also check

- General medical history including cardiac history and history of other neurological symptoms; medication (e.g. diuretics, TCAs)
- Psychiatric history—anxiety, depression, panic attacks?
- Family history—epilepsy, cardiac arrhythmia
- Substance abuse? Drugs or alcohol

Examination Usually normal by the time the patient is reviewed by a GP. Check temperature, respiratory rate, pulse, BP lying—and then standing for 3min. Listen to the heart. Record any neurological deficit.

Investigation 12-lead ECG (immediately or in <3d); consider checking capillary blood glucose if DM and/or FBC if any suggestion of bleeding.

Significant ECG changes[N]

- Any ventricular arrhythmia (including ventricular ectopics)
- Long QT (>450msec) or short QT (<350msec)—➲ p. 239
- Brugada syndrome (coved ST segment elevation >2mm in >1 of V1–V3 followed by a −ve T wave—may be transient)—➲ p. 239
- Ventricular pre-excitation (delta wave—slurring slow rise of the initial portion of the QRS complex—seen in WPW syndrome)—➲ p. 239
- Inappropriate significant bradycardia
- Left or right ventricular hypertrophy
- Abnormal T-wave inversion
- Pathological Q waves
- Sustained atrial arrhythmia
- Paced rhythm

Management[N] Treat any underlying cause identified.

Admit If any residual neurological deficit to exclude stroke.

Refer to cardiology for assessment in <24h if[N]*:*

- Any abnormalities on ECG
- Heart failure (history/clinical signs)
- Heart murmur
- New/unexplained breathlessness
- Transient loss of consciousness during exertion
- Family history of sudden cardiac death <40y and/or inherited heart condition

Consider referral if ≥65y and transient loss of consciousness without prodromal symptoms.

Refer urgently to neurology if[N]*:*

- Inexplicable gaps in recollection of events
- Prodromal déjà vu (or jamais vu)—➔ p. 547
- Tongue biting, unusual posturing or head turning to one side, and/or prolonged limb-jerking during the event (❗ brief seizure-like activity can be associated with simple faints)
- Confusion following the event

Epilepsy is *less* likely if prodromal symptoms are recognized by the patient and on previous occasions have been alleviated by sitting/lying; the episode was precipitated by prolonged standing; or there is sweating before or pallor during the episode.

Refer for routine cardiology review[N] If urgent referral is not indicated, no cause identified, and the episode was atypical for a simple faint, situational syncope, or orthostatic hypotension.

Vasovagal syncope (simple faint) Peripheral vasodilation, bradycardia, and venous pooling → postural hypotension. ♀ > ♂. Diagnose if no features suggesting an alternative diagnosis + any of the 3 'P's:

- Posture—prolonged standing before the event, or similar episodes in the past that have been prevented by lying down (*presyncope*)
- Provoking factors, e.g. pain or a medical procedure
- Prodromal symptoms, e.g. sweating, feeling warm/hot before the event

Situational syncope Provoked by straining during micturition or when opening bowels, coughing, blowing, or swallowing.

Management Reassure. No treatment is necessary.

Orthostatic (postural) hypotension ➔ p. 217

Hypoglycaemia Affects patients with DM taking insulin/oral hypoglycaemic agents. Produces autonomic changes (pallor, sweating, tachycardia) and behavioural changes (confusion, altered personality). If action is not taken to ↑ blood sugar, coma ± fitting ensues—➔ p. 324.

Epilepsy ➔ p. 546

Dizziness/giddiness ➔ p. 523

Hyperventilation and panic attacks ➔ p. 1081. ❗ Usually history is diagnostic but temporal lobe seizures may have similar symptoms.

Abnormal perceptions e.g. hallucinations—➔ p. 966

Further information

NICE (2010, updated 2014) Transient loss of consciousness ('blackouts') in over 16s. 🖱 www.nice.org.uk/guidance/cg109

Assessment of headache

Common presenting complaint. The skill lies in deciding which headaches are benign needing no intervention, and which require action.

History

- **Is there >1 type of headache?** Take a separate history for each
- **Time** When did the headaches start? How often do they happen? Is there a pattern (e.g. constant, episodic, daily)? How long do they last? Why is the patient consulting now? A headache diary over >8wk may be useful if longstanding headaches
- **Character** Nature/quality and site of pain. Associated symptoms, e.g. nausea/vomiting, visual or neurological symptoms
- **Cause** Predisposing and/or trigger factors; aggravating and/or relieving factors; relationship to menstrual cycle; family history
- **Response** Details of medication used (type, dose, frequency, timing). What does the patient do, e.g. can the patient continue work?
- **Health between attacks** Other medical history. Well between attacks?
- **Patient's anxieties and concerns**

Examination *In acute, severe headache*, examine for fever and purpuric skin rash. *In all cases*, check BP, brief neurological examination including fundi, visual acuity, and gait, palpation of the temporal region/sinuses for tenderness, and examination of the neck. *In young children*, measure head circumference and plot on a centile chart.

Refer To an appropriate specialist team. *E* = Emergency same-day assessment; *U* = Urgent; *S* = Soon; *R* = Routine.

- Fever and worsening headache ± purpuric rash/meningism—*E*
- ↓ consciousness and/or papilloedema—*E*
- Thunderclap headache (reaching peak intensity in <5min)—*E*
- New-onset seizures—*E/U*
- Recent head injury (<3mo)—*E/U/S*
- Change in personality/new cognitive or neurological deficit (consider referral for direct access MRI/CT in adults; specialist assessment in <48h for children)—*U*
- New-onset, unexplained, persistent headache in a patient with a history of HIV/immunosuppression or cancer metastasizing to the brain—*U*
- Suspected acute glaucoma (for ophthalmology review)—*U*
- Suspected temporal arteritis (for temporal artery biopsy)—*U*
- Progressive headache, worsening over weeks, especially if associated with vomiting, drowsiness, coughing/sneezing, or exercise—*U/S*
- Atypical migraine aura ± headache, e.g. motor weakness, double vision, visual symptoms in just one eye, poor balance, ↓ consciousness—*U/S*
- Substantial change in quality of usual headaches—*U/S*
- Persistent headache for >1mo or very frequent headaches, despite treatment affecting usual functioning—*S/R*

Differential diagnosis and management Table 15.8. ↑ BP may cause acute or chronic headache. Direct treatment at the cause.

Table 15.8 Differential diagnosis of headache

	Cause	Features	Management
Acute new headache	Meningitis	Fever, photophobia, stiff neck, rash	IV/IM penicillin V and immediate admission (➲ p. 1056)
	Encephalitis	Fever, confusion, ↓ conscious level	Immediate admission (➲ p. 1056)
	Subarachnoid haemorrhage	'Thunder-clap' or very sudden-onset headache ± stiff neck	Immediate admission (➲ p. 535)
	Head injury	Bruising/injury; ↓ conscious level, periods lucidity, amnesia	Consider admission (➲ p. 1094)
	Self-limiting viral illness	Vary. Often associated with other symptoms, e.g. coryza, sore throat, low-grade fever	Paracetamol/NSAID—review if worsens or if not settling in 2–3d
	Sinusitis	Tender over sinuses ± history of URTI	➲ p. 920
	Dental caries	Facial pain ± tenderness	➲ p. 910
	Tropical illness	History of travel, fever	➲ p. 624
Acute recurrent headache	Migraine	Aura, visual disturbance, nausea/vomiting, triggers	➲ p. 528
	Cluster headache	Nightly pain in 1 eye for 2–3mo then pain free for >1y	➲ p. 530
	Exertional or coital headache	Suggested by history of association	NSAID or propranolol before attacks
	Trigeminal neuralgia	Intense stabbing pain lasting seconds in trigeminal nerve distribution	➲ p. 531
	Glaucoma	Red eye, haloes, ↓ visual acuity, pupil abnormality	➲ p. 954
Subacute headache	Giant cell arteritis	>50y, scalp tenderness, ↑ ESR, rarely ↓ visual acuity	➲ p. 498
Chronic headache	Tension-type headache	Band around the head, stress, low mood	➲ p. 530
	Cervicogenic headache	Unilateral or bilateral; band from neck to forehead; scalp tenderness	➲ p. 448
	Medication overuse	Rebound headache on stopping analgesics	➲ p. 531
	↑ intracranial pressure	Worse on waking/sneezing, ↓ pulse, ↑ BP, neurological signs	➲ p. 532
	Paget's disease	>40y, bowed tibia, ↑ alk phos	➲ p. 478

Migraine

Migraine affects 15% of the UK population. ♂:♀ ≈1:3. 1 in 3 experience significant disability. Caused by disturbance of cerebral blood flow under the influence of serotonin.

Aura Occurs with or without headache. Symptoms arise over ≥5min and last 5–60min before resolving completely. Diagnose if:

- Visual symptoms, e.g. flickering lights, spots, lines; partial loss of vision
- Sensory symptoms, e.g. numbness; paraesthesia, and/or
- Speech disturbance

Atypical aura Consider referral for further investigation if: motor weakness; double vision; visual symptoms affecting only one eye; poor balance; or ↓ level of consciousness

Migraine headache Moderate to severe unilateral or bilateral throbbing/pulsating headache that lasts 4–72h (1–72h in children) and prevents usual activities. May occur with or without aura and be associated with nausea/vomiting ± ↑ sensitivity to light/noise.

- Episodic Occurs on <15d/month
- Chronic Occurs on ≥15d/month over >3mo

History, examination, and differential diagnosis ➔ p. 526

Management of an acute attack[N] Combination therapy with:

- Triptan (e.g. sumatriptan 50–100mg po)—choice depends on cost. Not effective if taken before the headache develops. Stops 70–85% attacks. Start with lowest dose and ↑ as needed. If consistently ineffective try an alternative triptan. Consider nasal triptan as first line if aged 12–17y
- NSAID (e.g. naproxen 500mg bd) *or* paracetamol (1g qds) ± antiemetic (e.g. prochlorperazine 5mg, metoclopramide 10mg, or domperidone 10–20mg)—even if no nausea/vomiting

If oral preparations are ineffective/not tolerated, offer metoclopramide 10mg PR or buccal prochlorperazine 3–6mg and consider adding a non-oral NSAID (e.g. diclofenac 100mg PR) or triptan (e.g. sumatriptan 20mg nasal spray or 6mg sc).

⚠ Do not offer ergots or opioids for the acute treatment of migraine.

Treatment of recurrence within the same attack Repeat symptomatic treatments within their dose limitations—pre-emptively if recurrence is usual/expected. If using triptans, a 2nd dose may be effective, but repeated dosing can cause rebound headache. Naratriptan and eletriptan are associated with relatively low recurrence rates.

Management of chronic migraine Aims to control symptoms and minimize impact on the patient's life. Cure is not a realistic aim.

Trigger factors Half have a trigger for their migraine. *Consider*:

- Psychological factors Stress/relief of stress; anxiety/depression; extreme emotions, e.g. anger or grief
- Environmental factors Loud noise, bright/flickering lights, strong perfume, stuffy atmosphere, VDUs, strong winds, extreme heat/cold

- Food factors Lack of food/infrequent meals; foods containing monosodium glutamate, caffeine, and tyramine; specific foods, e.g. chocolate, citrus fruits, cheese; alcohol, especially red wine
- Sleep Overtiredness (physical/mental); changes in sleep patterns (e.g. late nights, weekend lie-in, shift work, holidays); long-distance travel
- Health factors Hormonal changes (e.g. monthly periods, COC pill, HRT, the menopause);↑ BP; toothache or pain in the eyes, sinuses, or neck; unaccustomed physical activity

Assessing severity Assessment scales, e.g. Migraine Disability Assessment Score (MIDAS—➔ p. 557) can be useful in assessing impact of symptoms on daily life and monitoring response to treatment.

General measures Reassure. Instruct about management of acute attacks. A diary can be used to identify trigger factors, assess headache frequency, severity, medication usage/overusage, and response to treatment. Avoid trigger factors where possible. Give advice on relaxation techniques and stress management. ❗ Do not offer COC to women with migraine, especially if aura (Box 21.3, ➔ p. 731).

Prophylaxis[N] Consider if ≥4 attacks/mo or severe attacks. ↓ attacks by ~50%. Try a drug for 2mo before deeming it ineffective. If effective, continue for 6mo then consider ↓ the dose slowly and stopping.

- 1st line Propranolol S/R 80–160 mg od/bd or topiramate 25–50mg od/bd—start at low dose and ↑ dose every 2–4wk; ❗ topiramate is teratogenic and may ↓ effectiveness of hormonal contraception
- 2nd line Gabapentin (up to 1200mg/d in divided doses) or acupuncture (up to 10 sessions over 5–8wk)
- 3rd line Consider referral—botulinum type A toxin may be helpful for patients who have chronic migraine, do not have medication overuse headache, and have not responded to ≥3 different prophylactic drugs[N]

Alternative therapies Riboflavin 400mg od may ↓ frequency/intensity of headaches[N]; feverfew 200mg/d—may ↓ symptoms after 6wk use[C].

Menstrual migraine[N] Suspect if migraine occurs from 2d before to 3d after start of period on at least 2 out of 3 consecutive months (use headache diary). If predictable menstrual-related migraine that does not respond to standard acute treatment, consider frovatriptan (2.5mg bd) or zolmitriptan (2.5 mg bd/tds) on the days that migraine is expected.

Referral ➔ p. 526

> ❗ >1 type of headache may be present—50% migraine sufferers develop tension-type headache. Consider each separately.

Further information

NICE (2012, updated 2015) Headaches in young people and adults. 🖱 www.nice.org.uk/guidance/cg150

NICE (2012) Migraine (chronic)—botulinum toxin type A. 🖱 www.nice.org.uk/guidance/ta260

Patient information and support

Migraine Action Association ☎ 08456 011033 🖱 www.migraine.org.uk

Migraine Trust ☎ 020 7631 6970 🖱 www.migrainetrust.org

Other headaches and facial pain

Headache: assessment and differential diagnosis ➔ p. 526

Migraine ➔ p. 528

Chronic daily headache Prevalence 4%. Defined as any headache that occurs >15d/mo. *Common causes*: tension-type headache, cervicogenic headache (➔ p. 448), medication overuse headache, migraine, errors of refraction (usually headache is mild, frontal, in the eyes themselves, and absent on waking). Treat the cause (may be >1).

Tension-type headache^{N} Associated with stress and anxiety and/or functional or structural abnormalities of the head or neck. Prevalence ≈2%. ♀:♂ ≈2:1. Symptoms begin aged <10y in 15% patients. Prevalence ↓ with age. Family history of similar headaches is common (40%) but twin studies do not suggest a genetic basis. Distinguish between episodic and chronic tension-type headache:

- Episodic Defined as headache lasting 30min–7d and occurring <180d/y (<15d/mo)
- Chronic Headaches on ≥15d/mo (≤180d/y) for ≥3mo

In both cases pain:

- Is bilateral, pressing, and/or tightening in quality
- Of mild/moderate intensity
- Does not prohibit activities
- Is not associated with vomiting
- Is not aggravated by physical activity

Management Reassure no serious underlying pathology.

- Acute management—simple analgesia, e.g. paracetamol, ibuprofen. Avoid codeine-containing preparations and other opioids. Advise short-term use of analgesics only. At risk of medication overuse headache and/or opioid dependence if analgesics are used long term
- Try measures to alleviate stress—relaxation; massage; yoga; exercise
- Treat musculoskeletal symptoms with physiotherapy
- Consider referral for prophylaxis with acupuncture (if available)—up to 10 sessions over 5–8wk

Cluster headaches Extremely painful headaches focused around one eye with associated autonomic symptoms on that side (drooping eyelid, constricted pupil, red watery eye, runny or blocked nose, forehead sweating). Rare <20y of age. ♂:♀ ≈6:1. More common in smokers. Pain lasts 15–180min and occurs from 1× every 2d to 8×/d. Recurrences affect the same side. Onset is often predictable (1–2h after falling asleep; after alcohol). 2 patterns:

- Episodic—remissions of >1mo
- Chronic—remissions of <1mo in a 12mo period

Management Refer for specialist advice/neuroimaging for first bout of cluster headache. *Drug treatments*:

- Acute attack 100% oxygen (>12L/min) for 10–20min; $5HT_1$ agonists, e.g. sumatriptan (6mg sc or 20mg intranasal)—stops 75% in <15min
- Prophylaxis Consider verapamil 80mg tds/qds if attacks are frequent (needs ECG monitoring—seek specialist advice if unfamiliar with use). More effective if initiated early at the start of a new cluster. Refer for specialist advice if no response to verapamil

Medication overuse (analgesic) headache[N] Persistent headache in patients with other causes of pain who are overusing analgesics. Affects 1 in 50 adults; ♀:♂ ≈5:1. Consider if headache develops/worsens when taking analgesic medication for ≥3mo. Implicated drugs include:

- Triptans, opioids, ergots, or combination analgesics on ≥10d/month
- Paracetamol, aspirin, or NSAID on ≥15d/month

Management Explain the condition to the patient. Advise stopping overused medication abruptly for at least 1mo. Provide follow-up and support over 4–8wk; warn that symptoms may worsen initially (day 3–7) before improving. Review treatment of underlying problems (e.g. migraine, chronic musculoskeletal pain). Consider specialist referral if taking strong opioids or recurrent, failed attempts to stop medication overuse.

Facial pain Treat the cause. *Common causes include*: trigeminal neuralgia; temporomandibular joint disorders; dental disorders; sinusitis; migrainous neuralgia; shingles and post-herpetic neuralgia. No cause is found in many patients—it is then termed atypical facial pain. Atypical facial pain may respond to simple analgesia with paracetamol or NSAID. If this fails, try nerve painkillers, e.g. amitriptyline 10–75mg nocte. If troublesome symptoms, refer to ENT, maxillofacial surgery, or neurology.

Trigeminal neuralgia Paroxysms of intense stabbing, burning, or 'electric shock'-type pain lasting seconds to minutes in the trigeminal (V) nerve distribution; 96% unilateral. Mandibular/maxillary > ophthalmic division. Between attacks there are no symptoms. Frequency of attacks ranges from hundreds/day to remissions lasting years. Pain may be provoked by movement of the face (talking, eating, laughing) or touching the skin (shaving, washing). Can occur at any age but more common >50y. ♀ > ♂. Unknown cause but associated with MS in younger patients.

Management[N] Spontaneous remission may occur. The drug of choice for treatment of trigeminal neuralgia pain is carbamazepine (unlicensed indication). Start at low dose, e.g. 100mg od/bd and ↑ dose over weeks until symptoms are controlled. Usual dose ≈200–400mg tds. Often poorly tolerated. Oxcarbazepine is an alternative. If no response in <8wk, refer early for specialist assessment and advice on pain control.

Refer to neurology if: <50y; neurological deficit between attacks; treatment with carbamazepine/oxcarbazepine fails—specialist options include other neuropathic painkillers (amitriptyline, gabapentin, pregabalin, duloxetine), lamotrigine, baclofen, phenytoin, or surgical intervention.

Referral ➔ p. 526

Further information

NICE (2012, updated 2015) Headaches in young people and adults. 🖱 www.nice.org.uk/guidance/cg150
NICE (2013, updated 2019) Neuropathic pain. 🖱 www.nice.org.uk/guidance/cg173

Patient information and support

Organisation for the Understanding of Cluster Headaches (OUCH UK)
☎ 01646 651 979 🖱 www.ouchuk.org
Trigeminal Neuralgia Association UK ☎ 01883 370214 🖱 www.tna.org.uk

Raised intracranial pressure

Raised intracranial pressure (↑ ICP) usually presents with increasing headache associated with drowsiness, listlessness, vomiting, focal neurology, and/or seizures. *Causes include*: 1° or 2° tumours, head injury, intracranial haemorrhage, hydrocephalus, meningitis, encephalitis, brain abscess, and cerebral oedema (2° to tumour, trauma, infection, ischaemia).

Clinical features of ↑ ICP ⚠ If suspected, admit as an emergency.

- Drowsiness
- ↓ conscious level
- Irritability
- VI nerve palsy
- Papilloedema
- Dropping pulse
- Rising BP
- Focal neurological signs—due to underlying pathology
- Pupil changes—constriction then dilatation

Benign intracranial hypertension Symptoms/signs of a SOL but none is found. Usually occurs in young, obese women. Cause unknown. Treated with repeat lumbar puncture, ventriculoperitoneal shunt, diuretics, or dexamethasone. Usually resolves—but 10% recur later.

Brain abscess May be single or multiple. Organisms reach the brain via the bloodstream, direct implantation, or local extension from adjacent sites (e.g. sinusitis). Present with ↑ ICP, focal neurological signs, systemic effects of infection, and/or local effects due to the cause. Usually, features develop over 2–3wk—occasionally more slowly; if immunosuppressed, onset is rapid. If suspected, admit as an emergency. Treatment is with IV antibiotics ± surgical drainage. Mortality is 20–30%. 50% of survivors have long-term neurological deficit; 30% epilepsy.

Intracranial tumours

- **1° tumours** 70%. Classified by whether benign/malignant and cell type. Glioma is an umbrella term meaning tumour of nervous system origin. *Common subtypes*: astrocytoma, oligodendroglioma, glioblastoma multiforme, ependymoma. Tumours of the meninges (*meningioma*) and cerebral blood vessels (*cerebellar haemangioblastoma*) can also occur
- **2° tumours** 30%—usually from carcinoma of breast, lung, or melanoma—in 50% tumours are multiple

Presentation ❗ <1% of patients with headache have a brain tumour.

- **↑ ICP** 23–50% have papilloedema at presentation; headache 25–35%
- **Seizures** 25–30%. Suspect in all adults who have a first seizure—especially if focal or with localizing aura. Refer for urgent assessment[N]
- **Evolving focal neurology** Depends on the site. >50% have focal neurology at presentation. Frontal lobe lesions tend to present late
- **False localizing signs** Caused by ↑ ICP. VI nerve palsy (causing double vision) is most common due to its long intracranial course
- **Subtle personality change** 16–20% at presentation—irritability, lack of application, lack of initiative, socially inappropriate behaviour
- **Local effects** Skull base masses, proptosis, epistaxis

Differential diagnosis Stroke, MS, head injury, vasculitis, encephalitis, Todd's palsy (➔ p. 546), metabolic/electrolyte disturbance, other causes of SOL (e.g. abscess, subdural haematoma, cyst).

Prognosis Gliomas: all have ~20% 5y survival. Depending on site, meningiomas and haemangioblastomas have better prognosis.

Driving Inform the DVLA and seek specialist advice.

⚠ Referral guidelines for suspected brain tumour[N]

- Adults If available, urgent direct access brain MRI (or CT scan if MRI is contraindicated), to be performed in <2wk, if progressive, sub-acute loss of central neurological function (including unexplained personality change/cognitive deficit and/or cranial nerve palsy). If direct access MRI/CT is not available, refer to be reviewed by a neurologist in <2wk
- Children and young people Very urgent specialist paediatric referral (for an appointment in <48h) if newly abnormal cerebellar or other central neurological function

Other reasons to refer for neurological assessment[N]

E = Emergency same-day assessment; *U* = Urgent; *S* = Soon; *R* = Routine

- Papilloedema—*E*
- New-onset seizures—*E/U*
- Unexplained transient loss of consciousness (➔ p. 524)—*U*
- New-onset, unexplained, persistent headache in a patient with a history of HIV/immunosuppression or cancer metastasizing to the brain—*U*
- Progressive headache, worsening over weeks, especially if associated with vomiting, drowsiness, coughing/sneezing, or exercise—*U/S*
- Atypical migraine aura ± headache, e.g. motor weakness, double vision, visual symptoms in just one eye, poor balance, ↓ consciousness—*U/S*
- Substantial change in quality of usual headaches—*U/S*
- Persistent headache for >1mo or very frequent headaches affecting usual functioning—*S/R*

Hydrocephalus Dilatation of the cerebral ventricles and accumulation of CSF. May be:

- Communicating Due to ↑ production or ↓ reabsorption of CSF. *Causes*: post-meningitis; SAH (80% develop some degree of hydrocephalus); trauma; neoplastic infiltration in the subarachnoid space
- Non-communicating CSF flow is blocked due to an obstruction within the ventricles. Due to congenital malformations, tumour, brain abscess, SAH, meningeal scarring due to meningitis, or cranial trauma

Presentation and management

- Infants and children—➔ p. 878
- Adults Presents with ↑ ICP. Refer for urgent neurological assessment

❗ All patients with a CSF shunt should have pneumococcal vaccination.

Further information

NICE (2012, updated 2015) Headaches in young people and adults. 🖱 www.nice.org.uk/guidance/cg150
NICE (2012, updated 2018) Epilepsies: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg137
NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Information and support for patients

Brain & Spine Foundation ☎ 0808 808 1000 🖱 www.brainandspine.org.uk

Acute stroke and intracranial bleeds

Acute stroke Clinical syndrome typified by rapidly developing signs of focal or global disturbance of cerebral functions, lasting >24h or leading to death, with no apparent causes other than of vascular origin. Common and devastating condition—most common cause of adult disability in the UK. Half all strokes occur in people >70y. *Causes*:

- Cerebral infarction (≈70%). Atherothrombotic occlusion or embolism. *Sources of embolism*: left atrium (AF) or left ventricle (MI or heart failure). Ischaemia causes direct injury from lack of blood supply
- Intracerebral or subarachnoid haemorrhage (≈19%). Haemorrhage causes direct neuronal injury and pressure exerted by the blood results in adjacent ischaemia
- Rare causes Sudden ↓ BP, vasculitis, venous-sinus thrombosis, carotid artery dissection

Risk factors Age, ↑ BP, DM, AF, previous stroke/TIA, MI, artificial heart valves, hyperviscosity syndromes, smoking, alcohol, obesity, low physical activity.

Presentation Sudden onset of CNS symptoms, or stepwise progression of symptoms over hours or days (Box 15.1).

Assessment Check conscious level—may be ↓ or normal; record neurological signs (including dysphagia and incontinence); BP; heart rate and rhythm; heart murmurs; carotid bruits; systemic signs of infection or neoplasm. Screen for hypoglycaemia with a capillary blood glucose.

Acute management If stroke is suspected, admit directly to hospital by emergency ambulance. Thrombolysis early after stroke results in better outcomes, so do not delay referral until the patient is seen. Treatment of stroke in a stroke unit ↓ mortality and morbidity[G]. ⚠ Do not give aspirin prior to admission.

Differential diagnosis Decompensation after recovery from previous stroke (e.g. due to infection, metabolic disorder); SOL (1° or 2° cerebral neoplasm, cerebral abscess); trauma (subdural haematoma, traumatic brain injury); epileptic seizure; migraine; MS.

Driving and stroke/TIA Group 1 licence—stop driving for 1mo. Do not inform the DVLA unless residual deficit after 1mo. Group 2 licence—stop driving and inform the DVLA; relicensing is considered after 1y.

Secondary prevention of stroke ➔ p. 536

Transient ischaemic attack (TIA) History is as for stroke but there is full recovery in <24h. Patients who have a TIA have a 20% risk of stroke in the following month with highest risk in the first 72h.

Management If ongoing symptoms at the time of assessment, admit as for stroke. Otherwise give aspirin 300mg stat and arrange assessment by a specialized neurovascular assessment clinic in <24h[G].

Amaurosis fugax Form of TIA due to emboli passing through the retina. Transient loss of vision 'like a curtain'. Manage as for TIA.

Box 15.1 The FAST test

- Face—has the patient's face fallen on one side? Can he/she smile?
- Arms—can the patient raise both arms and keep them there?
- Speech—is the patient's speech slurred?
- Time—to call for an emergency ambulance

❗ The FAST test does not detect all strokes (e.g. sudden loss of vision, cerebellar symptoms). If any sudden-onset neurological symptoms without any other clear cause, treat as if stroke.

Subarachnoid haemorrhage (SAH) Spontaneous bleeding into the subarachnoid space. *Incidence*: 15/100,000. ♀ > ♂. Peak age 35–65y. High mortality. *Causes*: unknown (15%); rupture of congenital berry aneurysm (70%); AV malformation (15%). *Risk factors*: smoking, alcohol, ↑ BP, less common pre-menopause. Berry aneurysms may run in families and are associated with polycystic kidneys, coarctation of the aorta, and Ehlers–Danlos syndrome.

Presentation Typically sudden devastating headache ('thunderclap headache')—often occipital. Rarely (6%) preceded by a 'sentinel headache' representing a small leak ahead of the larger bleed. Vomiting/collapse with loss of consciousness ± fitting ± focal neurology follow. May be nothing to find initially on examination. Neck stiffness takes ~6h to develop. In later stages, may find ↓ level of consciousness, papilloedema, retinal or other intraocular haemorrhages, ± focal neurology.

Action If suspected admit immediately as a medical emergency. Only 1 in 4 admitted have an SAH. In most, no cause for the headache is found.

Subdural and extradural haemorrhage Bleeding either between the dura and bone of the skull (*extradural haemorrhage*), or from the bridging veins between cortex and venous sinuses, resulting in accumulation of blood between dura and arachnoid (*subdural haemorrhage*). *Causes*: trauma (may be trivial); idiopathic. *Risk factors*: Age, alcohol, falls, epilepsy, anticoagulant therapy.

Presentation Often insidious and history may go back several weeks:

- ↓ level of consciousness after head injury that initially produced no loss of consciousness; the 'lucid' interval may last from hours to days
- Fluctuation of conscious level (35% with subdural)
- Physical/intellectual slowing, unsteadiness on feet, confusion/personality change, and/or sleepiness
- Worsening headache ± symptoms of ↑ ICP (e.g. vomiting)
- Slowly evolving stroke (e.g. hemiparesis)

Differential diagnosis Stroke, cerebral tumour, dementia.

Action If suspected, admit as a medical emergency for further investigation. Evacuation of clot is possible even in very elderly patients and often results in full recovery. Outlook is less good if coma preoperative.

Further information

NICE (2019) Stroke and transient ischaemic attack in over 16s: diagnosis and initial management. 🖱 https://www.nice.org.uk/guidance/ng128

Prevention of stroke

Lifestyle measures for all patients

- Healthy diet (oily fish, low cholesterol, ↑ fruit and vegetables, ↓ salt) and, if obese, aim to ↓ weight until BMI <25 kg/m^2
- ↓ alcohol to ≤14u/wk over ≥3d, ↑ exercise, stop smoking (➔ p. 156)
- Treat any underlying DM ➔ p. 314

Primary prevention For patients with *no* history of stroke/TIA.

Cardiovascular risk reduction ➔ p. 214

- **Hyperlipidaemia** A 22% ↓ in cholesterol using a statin → a 30% ↓ in stroke in individuals with no past history of stroke/TIA. Treat if patients meet criteria for cardiovascular disease prevention (➔ p. 214)
- **Hypertension** A 5–6mmHg ↓ in BP reduces risk by >30% (➔ p. 218)

Anticoagulation Consider anticoagulation to ↓ stroke risk if:

- **Potential causes of cardiac thromboembolism** e.g. rheumatic mitral valve disease, prosthetic heart valves, dilated cardiomyopathy, or AF associated with valvular heart disease or prosthesis
- **Non-valvular AF** The CHA_2DS_2-VASc score (Table 15.9) is used to predict stroke risk and need for anticoagulation. Weigh risk of thromboembolism against risk of bleeding. The Keele University decision support tool may be useful: 🖱 https://www.anticoagulation-dst.co.uk

Choice of anticoagulant depends on cost, product indications/limitations, and patient preference. 💣 Anticoagulation for paroxysmal AF for 1° prevention is controversial—consider if any episodes lasting >30s.

After stroke Involve carers/families.

- Monitor and reassess frequently. Continue regular follow-up to monitor 2° prevention when specialist services have finished
- Refer for specialist rehabilitation if there is any deterioration in function. Aids/appliances/benefits can help—➔ p. 198 and ➔ p. 108
- Provide information about stroke and details of support organizations, verbal/written information about medicines and help with packaging, e.g. non-childproof tops, drug administration aid
- Screen both patients and carers for depression—➔ p. 173

Secondary stroke prevention[G] Patients with a history of stroke/TIA have a 26% risk of recurrent stroke within 5y.

- **Obstructive sleep apnoea** Independent risk factor for stroke. Screen all patients and refer for specialist assessment if +ve screen—➔ p. 308
- **Anticoagulation** Provide long-term anticoagulation for all patients with a history of non-haemorrhagic stroke/TIA + ongoing or paroxysmal valvular or non-valvular AF, or any other potential cause of cardiac thromboembolism. Choice of agent depends on cost, product indications/limitations, and patient choice. For warfarin, target INR = 2.5 (range 2–3) with 72% of readings within target range
- **Antiplatelet drugs** If non-haemorrhagic stroke/TIA and *not* taking an anticoagulant, offer clopidogrel 75mg daily long term. Combined aspirin 75mg od + dipyridamole S/R 200mg bd is an alternative if intolerant to clopidogrel—or either drug alone if combination is not tolerated
- **Hypertension management** ➔ p. 219. Start treatment <2wk after stroke/TIA. Aim to keep systolic BP <130mmHg unless carotid artery stenosis when target systolic BP is 140–150mmHg

- Lipid lowering ➔ p. 222. Treat all patients with a history of stroke/TIA with atorvastatin 80mg od as first line, regardless of baseline cholesterol. Aim to ↓ non-HDL cholesterol by >40% from baseline
- Carotid stenosis Patients with history of non-disabling stroke should be referred for consideration of surgery if 50–99% stenosis
- Antiphospholipid syndrome Consider screening with IgG/IgM anticardiolipin ELISA and lupus anticoagulant if unexplained ischaemic stroke/TIA, particularly if aged <50y, any autoimmune rheumatic disease, past history DVT/PE, recurrent 1st- or any 2nd/3rd-trimester pregnancy loss

Table 15.9 CHA$_2$DS$_2$-VASc score

	Condition	Points	Score
C	Congestive heart failure	1	0 *Low risk*—no oral anticoagulation 1 *Moderate risk* (1.3%/y)—oral anticoagulation depending on patient preference ≥2 *High risk*—oral anticoagulation unless contraindicated *Target INR for warfarin*: 2–3 + in target range for >72% of time
H	Hypertension	1	
A	Age >75y	2	
A	Age 65–74y	1	
D	DM	1	
S	Female[a]	1	
S	Prior stroke/TIA	2	
VASc	Vascular disease, e.g. MI, peripheral arterial disease, aortic plaque	2	

[a] For women <65y with no other risk factors, CHA$_2$DS$_2$-VASc score = 0

! In all cases weigh benefit of treatment against potential harms. The *HAS-BLED* score may help with decision-making:

- Hypertension (uncontrolled, systolic >160mmHg)—1 point
- Abnormal liver function (cirrhosis, bilirubin >2× or ALT/AST/alk phos >3× upper limit of normal)—1 point
- Abnormal renal function (dialysis, Cr >200micromol/L)—1 point
- Stroke history—1 point
- Prior major bleed or predisposition to bleeding—1 point
- Labile INR (<60% of the time in therapeutic range)—1 point
- Elderly (age ≥65y)—1 point
- Drugs predisposing to bleeding (e.g. antiplatelet agents, NSAIDs)—1 point
- Alcohol use—1 point

A score ≥3 indicates ↑ 1y bleed risk on anticoagulation sufficient to justify caution before prescribing anticoagulant and/or more regular review.

Source: data for CHA$_2$DS$_2$-VASc score from Lip et al. *Chest* 2010;**137**(2):263–72; data for HAS-BLED score from Pisters et al. *Chest* 2010;**138**(5):1093–100.

Further information

Royal College of Physicians (2016) National stroke guidelines. 🖱 www.rcplondon.ac.uk/guidelines-policy/stroke-guidelines

Patient information and support

Different Strokes ☎ 0345 130 7172 🖱 www.differentstrokes.co.uk
Stroke Association ☎ 0303 3033 100 🖱 www.stroke.org.uk

Parkinsonism and Parkinson's disease

Parkinsonism Syndrome of:
- Tremor Coarse tremor, most marked at rest, 'pill-rolling'
- Rigidity Limbs resist passive extension throughout movement—*lead-pipe rigidity*—and juddering on passive extension of the forearm or pronation/supination—*cogwheel rigidity*
- Difficulty in initiating movement
- Slowness of movement *Mask-like* or expressionless face, ↓ blink rate, ↓ fidgeting, ↓ peristalsis
- Abnormal gait Small steps ('*shuffling gait*') and flexed posture as if hurrying to keep up with feet ('*festinant gait*')
- Micrographia Small handwriting

Causes
- Parkinson's disease (PD)
- Other neurodegenerative diseases, e.g. Alzheimer's disease, multisystem atrophy
- Following encephalitis
- Drugs, e.g. haloperidol, chlorpromazine, metoclopramide
- Toxins, e.g. CO poisoning
- Trauma
- Normal pressure hydrocephalus

Treatment of drug-induced parkinsonism If possible, stop the implicated drug. If on an antipsychotic for schizophrenia, do not stop treatment, but add an antimuscarinic (e.g. procyclidine 2.5mg tds). Consider switching to an atypical antipsychotic drug—take specialist advice.

Steel–Richardson–Olszewski syndrome Parkinsonism accompanied by absent vertical gaze and dementia. Due to progressive supranuclear palsy. Treatment is supportive.

Parkinson's disease (PD) Incurable, progressive, degenerative disease affecting the dopaminergic neurons of the substantia nigra in the brainstem resulting in deficiency of dopamine and relative excess of acetylcholine transmitters. *Cause*: unknown. *Lifetime risk*: 1 in 40. ♂ = ♀. *Peak age at onset*: ≈65y but 5–10% patients are diagnosed when <40y old. Prevalence ↑ with age. Presents with parkinsonian symptoms/signs.

Non-motor symptoms
- Neuropsychiatric Apathy, anxiety/depression, visual hallucinations, psychosis, dementia, pain, olfactory disturbance
- Sleep Excessive day time sleepiness, restless legs
- Autonomic Drooling, postural hypotension, hyperhidrosis, urinary dysfunction, dysphagia, weight ↓, constipation, sexual dysfunction

Management Aims to: ↓ symptoms and ↑ quality of life; ↓ rate of disease progression; and limit side effects of treatment.

Screening for depression ➔ p. 173

Referral Refer all patients to a specialist with an interest in PD for confirmation of diagnosis, advice on management, and to access a multidisciplinary specialist rehabilitation team.

Rehabilitation Liaise closely with the specialist rehabilitation team.
- General principles ➔ p. 196
- Specific issues ➔ p. 554

Driving Drivers holding both group 1 and group 2 licences must inform the DVLA. Driving may continue as long as safe vehicle control can be maintained. A licence may be issued subject to regular review.

Carers ➔ p. 200

Drug treatment of Parkinson's disease Rarely achieves complete control of symptoms; 5–10% respond poorly. Treatment for PD should be consultant initiated and is not started until symptoms cause significant disruption of daily activities. *Options*:

Dopamine receptor agonists e.g. bromocriptine, pergolide. Often used alone as 1st-line treatment. ↑ dose gradually according to response and tolerability. Withdraw gradually. Can also be used in association with levodopa to ↓ off times and motor impairment.

> ⚠ Bromocriptine, pergolide, cabergoline, and lisuride have been associated with pulmonary, retroperitoneal, and pericardial fibrosis.
> - Check CXR ± spirometry, ESR, and creatinine before starting
> - Monitor for dyspnoea, persistent cough, chest pain, cardiac failure, abdominal pain or tenderness

Levodopa (or L-dopa) Precursor of dopamine. ↑ dopamine levels within the substantia nigra. Start with low dose and ↑ in small steps—aim to keep final dose as low as possible and a compromise between ↑ mobility and dose-limiting side effects (involuntary movements, psychiatric effects). Optimum dose interval varies between individuals.

- Only effective for PD. Not effective for patients with parkinsonism due to other causes. Improves bradykinesia and rigidity > tremor
- Often given with a co-drug (carbidopa or benserazide) which prevents peripheral breakdown of levodopa to dopamine but does not cross the blood–brain barrier (e.g. Sinemet®, Madopar®)
- With time there is ↓ response and troublesome side effects appear:
  - On–off effect—fluctuation between periods of exaggerated involuntary movements and periods of immobility
  - End-of-dose effect—duration of benefit after each dose reduces
  - Abnormal involuntary movements ↑

Other drugs

- Monoamine oxidase B inhibition e.g. selegiline, rasagiline. Used in severe PD in conjunction with levodopa to ↓ end-of-dose effect. Early use may postpone onset of treatment with levodopa
- Amantadine Improves bradykinesia, dyskinesias, tremor, and rigidity. Introduce and withdraw slowly
- Inhibition of enzymatic breakdown of dopamine, e.g. entacapone, tolcapone. For patients suffering from end-of-dose effect

Surgery A few patients benefit from 'deep brain stimulation' (DBS).

Further information

NICE (2017) Parkinson's disease in adults 🖱 www.nice.org.uk/guidance/ng71

Patient advice and support

Parkinson's Disease Society ☎ 0808 800 0303 🖱 www.parkinsons.org.uk

Multiple sclerosis

Chronic disabling neurological disease due to an autoimmune process of unknown cause. Characterized by formation of patches of demyelination ('plaques') throughout the brain and spinal cord. There is no peripheral nerve involvement. Most common neurological disorder of young adults with a lifetime risk of 1 in 1000. Peak age of onset is 20–40y. ♀:♂ ≈2:1.

Presentation Depends on the area of CNS affected. Take a careful history. Symptoms evolve over >24h, and persist over days/weeks before improving. Although usually presents with a single symptom, history may reveal other episodes that have gone unheralded. Isolated neurological deficits are never diagnostic. The hallmark of MS is a series of neurological deficits distributed in time and space not attributable to other causes.

Common features

- Visual symptoms ↓ vision, blurring or double vision; pain on eye movement (optic neuritis)
- Sensory/motor disturbance, e.g. numbness, tingling; Lhermitte's symptom (altered sensation travelling down back ± into limbs when bending the neck forwards); transverse myelitis (➔ p. 544)
- Problems with balance, unsteadiness, coordination, or clumsiness
- Problems with speech (e.g. slurring, slow)
- Pain (e.g. trigeminal neuralgia)
- Bladder/bowel problems (e.g. frequency, urgency, incontinence)
- Sexual dysfunction (e.g. erectile dysfunction)
- Non-specific symptoms—fatigue, depression, cognitive changes (e.g. loss of concentration, memory problems)

❗ Symptoms may be worsened by heat or exercise.

Prognosis

- **Benign MS** (10%) Retrospective diagnosis. The patient has a few mild attacks and then complete recovery. There is no deterioration over time and no permanent disability
- **Relapsing–remitting MS (RRMS)** 85% patients. Episodes of sudden ↑ in neurological symptoms or development of new neurological symptoms with virtually complete recovery after 4–6wk. With time remissions become less complete and residual disability accumulates
- **Secondary progressive MS (SPMS)** After ~15y, 65% of patients with relapsing–remitting disease begin a continuous downward progression which may also include acute relapses
- **Primary progressive MS (PPMS)** 10% patients. Steady progression from the outset with increasing disability

Management If suspected, exclude other causes[N]—check FBC, ESR/CRP, liver and renal function, Ca^{2+}, glucose/HbA1c, TFTs, vitamin B$_{12}$, and HIV serology. Refer to neurology for confirmation of diagnosis and support from the specialist neurological rehabilitation team. Urgency of referral depends on clinical circumstances.

Acute relapses[N] Refer for specialist review. First-line treatment is oral methylprednisolone 0.5g daily for 5d.

Disease-modifying drugs ↓ frequency and/or severity of relapses by ~30% and slow course of the disease (Box 15.2). All work through immune modulation ± anti-inflammatory effects. Prescription must be consultant led and monitored under the NHS risk sharing scheme.

Box 15.2 Disease-modifying drugs for MS

- Interferon beta
- Peginterferon-beta1a
- Glatiramer acetate
- Fingolimod
- Natalizumab
- Teriflunomide
- Dimethyl fumarate
- Alemtuzumab

Eligibility criteria Treatment should start as soon as possible after diagnosis—even if just a single clinical episode with radiological evidence of other lesions—if the patient is still able to walk with two crutches (Expanded Disability Status Scale (EDSS) of <7) and no evidence of progressive, non-relapsing MS. Patients experiencing a relapse with an EDSS of >7, may become eligible if recovery occurs.

Criteria to stop treatment

- After ≥6mo, there is no ↓ in frequency/severity of relapses compared to the pre-treatment phase
- Intolerable adverse effects
- EDSS falls to <6.5 (inability to walk) for >6mo due to MS
- Confirmed secondary progressive disease with ↑ in disability over a 12mo period in the absence of relapses

Depending on the reason for stopping, patients may be able to try another disease-modifying treatment.

Information[N] Offer all patients information about MS and local/national support groups.

- Encourage exercise; advise not to smoke (may ↑ MS progression)
- Discuss annual flu vaccination—eligible but may be contraindicated with some MS disease-modifying drugs and rarely triggers relapse
- For ♀ of childbearing age, inform that relapse rates ↓ in pregnancy; may ↑ for 3–4mo afterwards; but overall does not alter prognosis

Driving and MS All drivers must inform the DVLA. Driving can continue as long as safe vehicle control. Group 1 licence—may be restricted to cars with certain controls. Group 2 licence—annual review; licence is revoked if MS is progressive or sustained disability.

Management of symptoms and disability Liaise closely with the specialist neurological rehabilitation team.

- Screening for depression ➔ p. 173
- General principles of rehabilitation ➔ p. 196
- Common neurological rehabilitation problems ➔ p. 554

Further information

NICE (2014, updated 2019) Multiple sclerosis in adults: management. 🖱 www.nice.org.uk/guidance/cg186

Patient advice and support

MS Society ☎ 0808 800 8000 🖱 www.mssociety.org.uk

Motor neurone disease and CJD

Motor neurone disease (MND) Is a degenerative disorder of unknown cause affecting motor neurons in the spinal cord, brainstem and motor cortex. Prevalence in the UK is ~4.5/100,000; ♂:♀ ≈3:2. Peak age of onset ≈60y. 10% have a FH. There is *never* any sensory loss.

Patterns of disease There are 3 recognized patterns of MND:

- Amyotrophic lateral sclerosis (ALS) (50%) Combined lower motor neuron (LMN) wasting and upper motor neuron (UMN) hyperreflexia
- Progressive muscular atrophy (25%) Anterior horn cell lesions affecting distal before proximal muscles. Better prognosis than ALS
- Progressive bulbar palsy (25%) Loss of function of brainstem motor nuclei (LMN lesions) resulting in weakness of the tongue, muscles of chewing/swallowing, and facial muscles

Clinical picture Combination of progressive UMN and/or LMN signs affecting >1 limb or a limb and the bulbar muscles. May initially present as isolated/unexplained symptoms, including:

- Loss of dexterity, falls, or trips
- Speech/swallowing problems; tongue fasciculations
- Muscle problems, e.g. weakness, wasting, twitching, cramps, stiffness
- Breathing problems, e.g. shortness of breath
- Effects of ↓ respiratory function, e.g. fatigue, daytime sleepiness, early morning sleepiness or shortness of breath when lying down
- Cognitive features, e.g. behavioural changes, emotional lability, frontotemporal dementia

❗ MND *never* affects eye movements (cranial nerves III, IV, VI).

Management Refer to neurology for exclusion of other causes of symptoms and confirmation of diagnosis. MND is incurable and progressive. Death usually results from ventilatory failure 3–5y after diagnosis.

Prognostic factors Features at diagnosis associated with ↓ survival:

- Poor respiratory function
- Poor functioning in activities of daily living
- Older age
- Weight ↓
- Speech and swallowing problems (bulbar presentation)
- Shorter time from first developing symptoms to time of diagnosis

Disease-modifying therapy Should always be specialist initiated. Riluzole (50mg bd) is the only drug treatment licensed in the UK; it may extend life or time to mechanical ventilation for patients with ALS and slow functional decline. Monitoring of liver function is essential—monthly for the 1st 3 mo; then 3 monthly for 9mo; then annually thereafter.

Support Involve relevant agencies early, e.g. DN, social services, carer groups, self-help groups; apply for all relevant benefits (➔ pp. 104–9). Screen for depression (➔ p. 173). Regular review to help overcome any new problems encountered is helpful for patients and carers. Discuss the future and patients' wishes for the time when they become incapacitated with patients and carer(s).

Symptom control A multidisciplinary team approach is essential. Specific interventions:

- **Cramps** Quinine (first line); alternatives—baclofen, tizanidine, dantrolene, or gabapentin
- **Stiffness/spasticity** Baclofen, tizanidine, dantrolene, or gabapentin
- **Drooling** Propantheline 15–30mg tds po or amitriptyline 25–50mg tds; glycopyrronium bromide (if cognitive impairment); referral for botulinum toxin A
- **Dysphagia** Blend food, discuss nasogastric tubes/PEG (➔ p. 1018)
- **Depression** Common—reassess support, consider drug treatment and/or counselling
- **Respiratory failure** Discuss tracheostomy/ventilation—weigh pros and cons of prolongation of life versus prolongation of discomfort
- **Palliative care** ➔ p. 1011

Driving All drivers must inform the DVLA. Driving can continue as long as safe vehicle control. Group 1 licence—may be restricted to cars with certain controls. Group 2 licence—annual review; licence is revoked if MND is progressive or sustained disability.

Creutzfeldt–Jakob disease (CJD) (human spongiform encephalopathy) Fatal, degenerative 'prion' brain disease. *Types*:

- **Sporadic or classical** Most common form in the UK (≈50 cases/y). Rare <40y. Median duration of symptoms 3–4mo. *Cause*: unknown
- **Variant** Affects younger people than classical CJD and duration is longer, lasting a median of 14mo. *Cause*: transmitted by ingestion of nervous tissue in beef infected with bovine spongiform encephalitis or 'mad cow disease'. Compensation may be available to families
- **Familial prion disease** ~20–30 families in the UK are affected with a version of CJD passed from generation to generation in an autosomal dominant pattern. Median duration of symptoms from onset is 2–5y

Presentation Long incubation (>25y in some cases). Clinical features vary according to the areas of brain most affected but are always rapidly progressive. *Common features*: personality change; psychiatric symptoms; cognitive impairment; neurological deficits (sensory and motor deficits, ataxia); myoclonic jerks, chorea or dystonia; difficulty with communication, mobility, swallowing, and continence; coma and death.

Management There is no simple diagnostic test and often families feel frustrated by early misdiagnosis. Refer to neurologist if suspected. Treatment is supportive. *Palliative care*: ➔ p. 1011.

General principles of rehabilitation ➔ p. 196

Common neurological rehabilitation problems ➔ p. 554

Further information

NICE (2016, updated 2019) Motor neurone disease: assessment and management. 🖱 www.nice.org.uk/guidance/ng42

Patient advice and support

Brain & Spine Foundation ☎ 0808 808 1000 🖱 www.brainandspine.org.uk
Motor Neurone Disease Association ☎ 0808 802 6262 🖱 www.mndassociation.org

Spinal cord conditions

Spinal cord injury tends to affect young people, especially young men. It is devastating and the GP and primary care team are a vital part of the ongoing support network. *Causes*: trauma (42% falls; 37% RTAs), herniated disc, transverse myelitis, tumour, abscess.

Quadriplegia and tetraplegia Caused by spinal cord injury above the 1st thoracic vertebra. Usually results in paralysis of all four limbs, weakened breathing, and an inability to cough and clear the chest.

Paraplegia Occurs when the level of injury is below the 1st thoracic nerve. Disability can vary from the impairment of leg movement, to complete paralysis of the legs and abdomen up to the nipple line. Paraplegics have full use of their arms and hands.

Incomplete spinal cord injuries

- Anterior cord syndrome Damage is towards the front of the spinal cord, leaving the patient with loss or ↓ ability to sense pain, temperature, and touch sensations below the level of injury. Pressure and joint sensation may be preserved
- Central cord syndrome Damage is in the centre of the spinal cord. Typically results in loss of function in the arms, but preservation of some leg movement ± some control of bladder/bowel function
- Posterior cord syndrome Damage is towards the back of the spinal cord. Typically leaves patients with good muscle power, pain, and temperature sensation, but difficulty coordinating limb movements
- Brown-Séquard syndrome Damage is limited to 1 side of the spinal cord resulting in loss or ↓ movement on the injured side but preserved pain and temperature sensation, and normal movement on the uninjured side but loss or ↓ in pain and temperature sensation

Cauda equina lesion The spinal cord ends at L1/L2 at which point a bundle of nerves travels downwards through the lumbar and sacral vertebrae. Injury to these nerves causes partial or complete loss of movement and sensation. There may be some recovery of function with time.

Transverse myelitis Inflammation of the spinal cord at a single level. Symptoms develop rapidly over days/weeks and include limb weakness, sensory disturbance, bowel and bladder disturbance, back pain, and radicular pain. Recovery generally begins in <3mo but is not always complete. *Causes*:

- Idiopathic (thought to be autoimmune mechanism)
- Infection
- Vaccination
- Autoimmune disease, e.g. SLE, Sjögren's syndrome, sarcoidosis
- MS
- Malignancy
- Vascular, e.g. thrombosis of spinal arteries, vasculitis 2° to heroin misuse, spinal AV malformation

Management Depending on severity of symptoms, admit as an acute medical emergency or refer for urgent neurological opinion.

Syringomyelia Tubular cavities (syrinxes) form close to the central canal of the spinal cord. As the syrinx expands, it compresses nerves within the spinal cord. Most common in patients with previous spinal injury—although may be years before. Typically presents with wasting and weakness of hands and arms, and loss of temperature and pain sensation over trunk and arms (cape distribution). Refer to neurology if suspected.

General principles of rehabilitation ➲ p. 196

Common neurological rehabilitation problems ➲ p. 554

Specific problems associated with spinal cord injury

Autonomic dysreflexia (hyperreflexia) Reflex sympathetic overactivity causing flushing and ↑ BP which may be severe. Only occurs in patients with lesions above T5/6. Usually triggered by discomfort below the level of the lesion. Presentation is with pounding headache, sweating, flushing, or mottling above the level of the lesion.

Sit the patient up and remove any obvious cause, e.g. pain, bladder distension, constipation. Give GTN spray (1–2 puff sublingual) or nifedipine 5–10mg capsule broken sublingually. If not settling, admit to hospital.

Loss of temperature control Most people with complete spinal cord injuries do not sweat below the level of the injury and many quadriplegics cannot sweat above the injury (even though they may sweat due to autonomic dysreflexia). With loss of ability to sweat or vasoconstrict, careful control of environmental conditions is essential to avoid hypothermia or overheating. In hot weather, advise cooling with wet towels.

Infertility Many ♂ patients suffer infertility due to:

- Failure of ejaculation
- Retrograde ejaculation
- Thermal damage due to sitting in a wheelchair → poor quality sperm
- Chronic infection of prostate and seminal vesicles (common)

Refer for specialist advice.

Bowel/bladder function Both bladder and bowel function are reflex actions that we learn to override as children. If the lesion is above the level of this reflex pathway (T12 for bowel and T6 for bladder function) then automatic emptying will still occur when the bladder or bowel is full—though there is no control. If the lesion is below this level there is no emptying reflex. Bladder/bowel care programmes reflect this. Useful leaflets are available from the Spinal Injuries Association.

Spasticity ➲ p. 555

UTI ➲ p. 555

Pressure sores ➲ p. 585

Depression ➲ p. 173

Patient advice and support

Aspire 🖱 www.aspire.org.uk
Brain & Spine Foundation ☎ 0808 808 1000 🖱 www.brainandspine.org.uk
British Syringomyelia & Chiari Society 🖱 www.britishsyringomyelia-chiarisociety.org
Spinal Injuries Association ☎ 0800 980 0501 🖱 www.spinal.co.uk
Transverse Myelitis Association 🖱 www.myelitis.org.uk

Epilepsy

Epilepsy is a group of disorders in which fits or seizures occur as a result of spontaneous abnormal electrical discharge in any part of the brain. They take many forms but usually take the same pattern on each occasion for a given individual. Prevalence 5–10/1000. 5% of those >21y old having their first fit have cerebral pathology (10% if aged 45–55y).

Transient loss of consciousness ➲ p. 524

Epilepsy in children ➲ p. 876

Management of a fitting patient/status epilepticus ➲ p. 1080

Management after first fit 60% of adults who have one fit will never have another (90% if EEG is normal).

> ⚠ Refer *all* patients with a first suspected seizure for assessment by a neurologist with training and expertise in epilepsy urgently to exclude underlying causes (e.g. tumour) and receive clear guidance on medication, work, and driving[N].

Classification of seizure types Is important, as these have implications for management and prognosis:

- **Partial seizures** Limited to one area of the brain only. Termed 'simple' if no impairment of consciousness, and 'complex' if consciousness is impaired. Partial seizures may become generalized
- **Generalized seizures** Whole brain is involved. Consciousness is usually but not always impaired. 6 major types: tonic–clonic (grand mal); absence (petit mal); myoclonic; tonic; clonic; and atonic

❗ Some people have seizures that cannot be classified in this way.

Todd's palsy Focal CNS signs (e.g. hemiplegia) following an epileptic seizure. The patient seems to have had a stroke but recovers in <24h.

Causes of epilepsy in adults A cause is found in > two-thirds of people with epilepsy. The most common causes are:

- Cerebrovascular disease
- Cerebral tumours
- Genetic, congenital, or hereditary conditions
- Drugs, alcohol, or other toxic causes
- Head trauma (including surgery)
- Post-infective causes (e.g. meningitis, encephalitis)

Assessment Table 15.10

Screening for depression ➲ p. 173

Long-term management of epilepsy ➲ p. 548

Epilepsy and pregnancy ➲ p. 807

Driving and epilepsy ➲ p. 548

Mortality Death rate is ↑ ×2–3. Deaths are related to underlying condition, accidents, SUDEP, or status epilepticus.

Sudden unexplained death in epilepsy (SUDEP) Probably due to central respiratory arrest during a seizure. ↓ risk by optimizing seizure control and being aware of potential consequences of night seizures.

Table 15.10 Summary of points to cover during assessment

History		
Background	• Previous head injury • Alcohol/drug abuse • Meningitis or encephalitis	• Stroke • Febrile convulsions • Family history of epilepsy
Provoking factors	• Sleep deprivation • Alcohol withdrawal	• Flashing lights
Prodrome/aura	*Prodrome*—precedes fit. May be a change in mood or behaviour noticed by the patient or others *Aura*—part of the seizure that precedes other manifestations—odd sensations, e.g. déjà-vu (odd feeling of having experienced that time before), strange smells, rising abdominal sensation, flashing lights	
Features of the attack	*Eye witness report*: if available—colour of the patient, movement, length of fit, circumstances, after-effects *Memories of the patient*: of the event and first memories after the event, attack frequency, relationship to sleep, menses etc.	
Residual symptoms after the attack	• Bitten tongue • Incontinence of urine/faeces (not specific for epilepsy) • Confusion • Headache • Aching limbs or temporary weakness of limbs (Todd's palsy)	
Examination		
Neurological examination	• Fever, photophobia, neck stiffness or petechial rash? • Any residual focal neurology • Signs of ↑ ICP (➔ p. 532)	
General examination	• BP, heart sounds, heart rhythm and rate • Signs of systemic illness	
Investigations (first fit only)	• ECG • Capillary blood glucose • Blood: U&E, Cr, eGFR, LFTs, Ca^{2+}, FBC, ESR/CRP, HIV	
Differential diagnosis		
• Syncope (simple or situational) • Psychogenic non-epileptic attacks (pseudo-seizures) • Tics • Panic attack • Hypoglycaemia	• Normal phenomenon (e.g. déja-vu) • Cardiac disorders, e.g. arrhythmia, aortic stenosis, HOCM • TIA • Migrainous aura	

⚠ Avoid prescribing sodium valproate to girls of childbearing age. If essential, ensure a pregnancy prevention plan is in place and that information is provided about potential teratogenic effects both verbally and in writing.

Further information

NICE (2012, updated 2018) Epilepsies: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg137

Patient advice and support

Epilepsy Action ☎ 0808 800 5050. 🖱 www.epilepsy.org.uk

Management of epilepsy

Education Epilepsy is a diagnosis causing alarm and fear. How much does the patient/family understand about epilepsy? Acknowledge distress and answer questions. Provide information on:

- What epilepsy is and support available locally/nationally
- What to expect—fits are controlled with drugs in 80%; concordance with medication; when drug withdrawal may be considered
- What to do during an attack
- Work—inform employer. Do not work at heights or with/near dangerous machinery
- Avoiding risks—avoid cycling in traffic; only swim if lifeguard present

Driving Inform the DVLA and motor insurance company

- Single, unprovoked seizure Group 1—stop driving 6mo; group 2—licence restored if no seizures for >5y off medication
- Epilepsy or multiple unprovoked seizures Group 1—stop driving until fit-free for >1y; group 2—licence restored if seizure-free for >10y off antiepileptic medication

Drugs Table 15.11. Treatment usually starts after the 2nd seizure. Drug choice is a specialist decision. ❗ Patients can claim free prescriptions.

Withdrawal of drug therapy Consider if fit-free for 2–3y. Decision to stop *must* be the patient's. Balance problems/inconvenience of drug-taking against risks of fits. Refer to neurology for supervision of drug withdrawal. If adult with grand mal epilepsy, 59% stay fit free for 2y.

Seizure recurrence is more likely if generalized tonic–clonic seizures; myoclonic epilepsy or infantile spasms; taking >1 drug for epilepsy; ≥1 seizure after starting treatment; duration of treatment >10y; fit free <5y.

⚠ Patients must stop driving during medication withdrawal and for 6mo afterwards.

Other treatments

- Surgery Used for intractable partial seizures, hemiepilepsy, and epilepsy with focal EEG and/or radiological features
- Vagus nerve stimulation ↓ frequency of seizures for those refractory to antiepileptic medication but not suitable for respective surgery
- Ketogenic diet Effective in some patients with refractory epilepsy—take specialist advice

At annual review Review care plan. Record fits and precipitating causes; check drug concordance (frequency of repeat prescriptions) and side effects; if fit free >2y, discuss withdrawing medication. For ♀ of reproductive age give contraception (➔ p. 728)/pre-conception (➔ p. 807) advice.

Re-refer For review by a neurologist if:

- Control is poor or drugs are causing unacceptable side effects
- Seizures have continued despite medication for >2y or on 2 drugs
- Pointers to a previously unsuspected cause for the fits appear
- Concurrent illness (physical or psychiatric) complicates management
- For pre-conceptual advice or to discuss withdrawal of medication

Table 15.11 Commonly used drugs in epilepsy. Stress the importance of concordance. Principles of epilepsy medication: start at a low dose; ↑ dose until fits are controlled or side effects occur. Use monotherapy—2 drugs ↑ toxicity and side effects and offers no benefits over monotherapy for 90% patients. Prescribe by brand name—changing brand carries 10% risk of worsening of seizure control

		Ethosuximide	Sodium valproate[1]⚠	Carbamazepine	Lamotrigine	Clonazepam
Type of epilepsy	*Absence*	✓	✓		✓	✓
	Myoclonic		✓			✓
	Tonic–clonic		✓	✓	✓	
	Partial ± 2° generalized		✓	✓	✓	
Adult starting dose		500mg od	300mg bd	100–200mg od or bd	25mg od for 2wk[3]	1mg nocte for 4d
Incremental dose		250mg/d at weekly intervals	200mg/d at 3 day intervals	100mg/d at weekly intervals	From starting dose to 50mg od for 2wk then ↑ by 50mg/d at weekly intervals	↑ according to response over 2–4wk
Usual daily dose		1–1.5g od	500mg–1g bd	200–1200mg	100–200mg	4–8mg nocte
Common/important side effects		Blood dyscrasias[5], sedation, nausea, vomiting, dizziness, ataxia	⚠ Teratogenic[1] Pancreatitis, liver toxicity[4], blood dyscrasias[5], sedation, tremor, weight ↑, hair thinning, ankle swelling	Blood dyscrasias[5], rash, liver toxicity[4], nausea, sedation, diplopia, dizziness, fluid retention, ↓ sodium[2]	Blood dyscrasias[5], rash, fever, influenza-like symptoms, drowsiness or worsening of seizure control	Drowsiness/fatigue, amnesia/confusion/restlessness, muscle hypotonia, coordination problems, dependence and withdrawal

1. ⚠ Teratogenic—avoid prescribing to ♀ of childbearing age. If essential, ensure the ♀ is fully informed of potential risks and has reliable contraception—➔ p. 728.
2. Monitor U&E at regular review.
3. Starting dose is different if used in association with other epileptics—see *BNF*.
4. Warn about symptoms of liver disease. Check LFTs soon after starting and at review.
5. Check FBC if bruising, mouth ulcers, or symptoms of infection (sore throat, fevers).

Muscle disorders

Symptoms Muscle weakness, fatigability. Pain at rest suggests inflammation—pain on exercise, ischaemia, or metabolic myopathy.

Signs Look for associated systemic disease.

- Myotonia Delayed muscular relaxation after contraction, e.g. difficulty letting go after gripping something
- Local muscular tenderness or firm muscles May be due to infiltration of muscle with connective tissue or fat
- Fasciculation Spontaneous, irregular, and brief contractions of part of a muscle—suggests LMN disease, e.g. MND
- Lumps Tumours are rare—lumps may be due to tendon rupture, haematoma, or herniation of muscle through fascia

Muscular dystrophies Group of genetic disorders characterized by progressive degeneration and weakness of some muscle groups.

Dystrophia myotonica Autosomal dominant inheritance—abnormal *DMPK* gene on chromosome 19. Presents at any age. Symptoms vary from mild to severe and may include:

- Muscle symptoms Weakness and myotonia—particularly involves face, eyelids, jaw, neck, forearms/hands, lower legs/feet. Can affect speech and result in a lack of facial expression
- Respiratory symptoms Weakness of respiratory muscles → poor night-time sleep, daytime sleepiness, headaches, and difficulty waking; aspiration → recurrent chest infections
- Eye symptoms Cataract (may be the only problem) and ptosis
- Reproductive problems Infertility as a result of atrophy of the testes and problems in labour due to uterine muscle weakness
- Learning difficulty and behavioural problems
- Digestive symptoms— Swallowing difficulty, abdominal pain, constipation/diarrhoea, gallstones
- Cardiac arrhythmias Annual ECG is advisable
- Endocrine abnormalities e.g. DM
- Anaesthetic problems Pre-warn anaesthetist/surgeon prior to surgery

Prognosis is variable depending on severity of symptoms. Refer to confirm diagnosis, and for advice on management/genetic counselling.

Duchenne's muscular dystrophy Sex-linked recessive inheritance means almost always confined to boys. 30% of cases are due to spontaneous mutation. Investigation shows markedly ↑ CK (>40× normal). Presents typically at ~4y with progressively clumsy walking. Few survive to >20y old. Refer for confirmation of diagnosis and ongoing specialist support. Genetic counselling is important.

Patient information and support

Muscular Dystrophy Campaign ☎ 0800 652 6352 🖱 www.muscular-dystrophy.org

Myotonic Dystrophy Support Group ☎ 0115 987 0080 🖱 www.myotonicdystrophysupportgroup.org

Toxic myopathies Certain drugs can cause myopathy including:

- Alcohol
- Labetalol
- Cholesterol-lowering drugs (including the statins)
- Steroids
- Chloroquine
- Zidovudine
- Vincristine
- Ciclosporin
- Cocaine
- Heroin
- PCP

Management Stop the implicated drug immediately. If symptoms do not resolve, refer for confirmation of diagnosis and management advice.

Acquired myopathy of late onset Often a manifestation of systemic disease, e.g. thyroid disease (especially hyperthyroidism), carcinoma, Cushing's disease. Investigate to find the cause. Treat the cause if found, otherwise refer for further investigation.

Polymyositis Insidious, symmetrical, proximal muscle weakness due to muscle inflammation. Dysphagia, dysphonia, and/or respiratory muscle weakness may follow. 25% have a purple rash on cheeks, eyelids, and other sun-exposed areas (*dermatomyositis*) ± nail fold erythema. CK levels are ↑. Associated with malignancy in 10% of patients > 40y. Refer.

PoliomyelitisND

Acute polio *Spread*: droplet or faecal–oral. *Incubation*: 7d. Presents with 2d flu-like prodrome then fever, tachycardia, headache, vomiting, stiff neck, and unilateral tremor ('pre-paralytic stage'). 65% who experience the pre-paralytic stage go on to develop paralysis (myalgia, LMN signs ± respiratory failure). *Management*: supportive—admit to hospital. <10% of those developing paralysis die. Permanent disability may result.

Prevention

- 1° immunization in babies and children <10y 3 doses of the 6-part vaccine (DTaP/IPV/Hib/HepB) protecting against polio, diphtheria, whooping cough, tetanus, *Haemophilus influenza* type b, and hepatitis B, each 1mo apart—usually at 2mo, 3mo, and 4mo. If schedule is disrupted, resume where stopped
- Booster doses in children 1 dose of 4-part vaccine (DTaP/IPV) protecting against polio, diphtheria, whooping cough, and tetanus >3y after the 1° course (usually pre-school) and another dose of 3 part vaccine (Td/IPV) against tetanus, diphtheria and polio, 10y later (age 13–18y)
- 1° immunization in children >10y and adults 3 doses of 3-part vaccine (Td/IPV) each 1mo apart. Give booster doses after 3y and 10y
- Booster doses for travel Not required unless at special risk, e.g. travelling to endemic/epidemic area or healthcare workers. Boosters of Td/IPV are then given every 10y

Late effects of polio 20–30y after initial infection some patients develop new symptoms often triggered by a period of immobilization:

- ↑ muscle weakness and fatigue
- Pain in muscles and joints
- Respiratory difficulties (particularly in those who spent some time in an iron lung ventilator)—may present with symptoms relating to sleep

Once other causes are excluded, treatment is supportive.

Motor neurone disease ➔ p. 542

Myasthenia gravis/Lambert–Eaton syndrome ➔ p. 518

Other neurological syndromes

Von Recklinghausen's disease (type 1 neurofibromatosis; NF1) Autosomal dominant trait. *Criteria for diagnosis*: ≥2 of:

- ≥6 café-au-lait patches (flat, coffee-coloured patches of skin seen in 1st year of life, ↑ in number and size with age) >5mm (prepubertal) or >15mm (postpubertal)
- ≥2 neurofibromas:
  - Dermal neurofibromas—small violaceous skin nodules which appear after puberty
  - Nodular neurofibromas—subcutaneous, firm nodules arising from nerve trunks (may cause paraesthesiae if compressed) or a plexiform neurofibroma which appears as a large subcutaneous swelling
- Freckling in axilla, groin, neck base, and submammary area (women). Present by age 10y
- ≥2 Lisch nodules—nodules of the iris only visible with a slit lamp
- Distinctive bony abnormality specific to NF1, e.g. sphenoid dysplasia
- 1st-degree relative with NF1

Management Ongoing specialist management is essential.

Complications Affect 1 in 3 patients:

- Mild learning disability
- Short stature
- Macrocephaly
- Nerve root compression
- GI bleeding or obstruction
- Cystic bone lesion
- Scoliosis
- Pseudoarthrosis
- ↑ BP (6%)—due to renal artery stenosis or phaeochromocytoma
- Malignancy (5%)—optic glioma or sarcomatous change of neurofibroma
- Epilepsy (slight ↑)

Type 2 neurofibromatosis (NF2) Much rarer than type 1. Autosomal dominant inheritance.

Diagnosis One of:

- Bilateral vestibular schwannoma (acoustic neuroma—sensorineural hearing loss, vertigo ± tinnitus) *or*
- 1st-degree relative with NF2 *and either* a unilateral vestibular schwannoma *or* ≥1 neurofibroma, meningioma, glioma, schwannoma, or juvenile cataract

Management Screen at-risk patients with annual hearing tests. Once diagnosis is made, specialist neurosurgical management is needed.

Complications Schwannomas of other cranial nerves, dorsal nerve roots, or peripheral nerves; meningioma (45%); other gliomas (less common).

Ekbom syndrome (restless legs syndrome) The patient (who is usually in bed) is seized by an irresistible desire to move his/her legs in a repetitive way accompanied by an unpleasant sensation deep in the legs. Sleep disturbance is common, as is +ve FH. *Cause*: unknown.

Management

- Exclude drug causes—common culprits: β-blockers, H_2 antagonists, neuroleptics, lithium, TCAs, anticonvulsants

- Exclude peripheral neuropathy or ischaemic rest pain
- Iron deficiency (with or without anaemia) is associated in 1 in 3 sufferers, so check FBC and serum ferritin
- Also check: U&E, Cr, eGFR, fasting blood glucose, and TFTs
- Try non-drug measures first—reassurance, information, walking/stretching, warmth, relaxation exercises, massage
- Drugs—dopamine agonists are often effective, e.g. ropinirole, pramipexole
- Refer if severe symptoms or diagnosis is in doubt

Patient support

RLS-UK 🖱 https://www.rls-uk.org/

Wernicke's encephalopathy Thiamine deficiency causing nystagmus, ophthalmoplegia, and ataxia. Other eye signs, e.g. ptosis, abnormal pupillary reactions, and altered consciousness or confusion may also occur. Consider in any patient with symptoms and a history of alcohol misuse.

Management Refer for confirmation of diagnosis. Meanwhile start thiamine 200–300mg od po to prevent irreversible Korsakoff syndrome. In severe cases, admit as a medical emergency.

Korsakoff syndrome ↓ ability to acquire new memories. May follow Wernicke's encephalopathy and is due to thiamine deficiency. Confabulation to fill gaps in memory is a feature.

Gilles de la Tourette syndrome ➜ p. 889

Huntington's disease (chorea) Autosomal dominant trait. Testing can identify affected individuals before symptoms occur. Pre-conceptual and antenatal testing is available and should be offered to any couple with a family history on either the mother or the father's side. Presents with movement abnormalities (e.g. hemichorea and rigidity) and dementia. Memory is relatively spared compared to cognition. Refer to neurology for diagnosis and expert advice and to genetics for discussion of gene testing.

Friedreich's ataxia The most common inherited ataxia (autosomal recessive). Prevalence—1 in 50,000. Presents in adolescence with progressive gait and limb ataxia, loss of proprioception, pyramidal weakness, and dysarthria. Extra-neurological involvement includes hypertrophic cardiomyopathy (most patients) and DM (10%). Treatment is supportive. Most patients become chairbound within 15y and die in the 4th or 5th decade from cardiac or pulmonary complications.

Patient support

Ataxia UK ☎ 0845 644 0606 🖱 www.ataxia.org.uk

Neurological rehabilitation problems

General principles of rehabilitation ➔ p. 196

Fatigue Consider and treat factors that might be responsible:

- Depression
- Chronic pain
- Disturbed sleep
- Poor nutrition

Action Review support, diet, and medication; encourage graded aerobic exercise; consider a trial of amantadine 200mg/d to improve symptoms^{N}.

Depression and anxiety Common. Diagnosis can be difficult. Use NICE depression screening questions (➔ p. 173). Any positive response should prompt further assessment for depression (➔ p. 978). Talk about the impact of the illness on lifestyle. Jointly identify areas where +ve changes could be made, e.g. referral to day care to widen social contact. Consider referral for counselling or to a self-help/support group. Consider antidepressant medication and/or referral to psychiatric services.

Emotionalism If the patient cries (or laughs) with minimal provocation, consider emotionalism—impairment in the control of crying. Reassure.

Sexual and personal relationships Problems are common. Useful information sheets are available at 🖱 www.outsiders.org.uk

Communication problems Speech therapy assessment is vital. Consider support via dysphasia groups and communication aids, e.g. simple pointing board (take advice from speech therapy and OT).

Poor vision Refer to an optician in the first instance. If corrected vision is still poor, refer for ophthalmology review.

Respiratory infections Common. Treat with antibiotics unless in terminal stages of disease. Advise pneumococcal and influenza vaccination as appropriate (❗ Flu vaccine may be contraindicated for some patients with MS on disease-modifying therapy.)

Venous thromboembolism Common but clinically apparent in <5%. Ensure adequate hydration; encourage mobility. Consider use of compression stockings if immobile. Prophylactic anticoagulation does not improve outcome.

New symptoms or limitations Consider:

- Is it due to an unrelated disease (e.g. change in bowel habit in someone who has had a stroke might indicate bowel cancer)?
- Is it due to an incidental infection (e.g. UTI, chest infection)?
- Is it due to a relapse (e.g. acute relapse in MS, TIA, or further stroke)?
- Is it due to a side effect of treatment (e.g. acute confusion, involuntary movements, or the on–off effect in a patient with PD)?
- Is it part of a gradual progression (e.g. in MS, MND, brain tumour)?

Treat any cause of deterioration identified. If no cause is found, consider re-referring for specialist review and/or referring to the multidisciplinary rehabilitation team involved with the patient.

Motor impairment Aim to maintain physical independence:

- Involve physiotherapy ± OT. Often a task-oriented approach is used (e.g. learning how to dress). Can also supply/advise on aids and appliances, e.g. hook-and-loop fasteners, wheelchairs, adapted cutlery, etc.
- Refer for social services OT assessment if aids, equipment, or adaptations are needed for the home
- Refer for home care services as necessary
- Give information about driving and/or employment where appropriate

Spasticity ± muscle and joint contractures Treat with physiotherapy (usually involving exercise ± splinting) ± drugs. Antispasticity drugs include dantrolene (25mg od), baclofen (5mg tds or rarely through a pump), tizanidine (2mg od), and gabapentin (300mg–2.4g daily in divided doses. Botulinum toxin can be directed at specific muscles. Refer via the specialist rehabilitation team.

Pain Most pain arises from ↓ mobility. *Other causes include*: pre-morbid disease (e.g. osteoarthritis); central pain due to neurological damage; and neuropathic pain. Chronic pain, especially central pain, may respond to TCAs. Peripheral pain may respond to simple analgesia ± physiotherapy. Other options are TENS and local joint injection. A cannabinoid is now available as an oromucosal spray (Sativex®) for relief of pain/muscle spasm in MS on specialist prescription only. Refer patients with intractable pain to specialist pain clinics. ⚠ Avoid long-term opioids or benzodiazepines.

Bladder problems

- UTI If suspected check urine dipstick ± send MSU for M,C&S and start antibiotics. If >3 proven UTIs in 1y refer to specialist incontinence service or urology for further assessment
- Incontinence ➔ p. 424. If urgency, modify environment, e.g. provide commode; try anticholinergic, e.g. tolterodine 2mg bd or oxybutinin 5mg tds. If not settling, refer for specialist assessment
- Nocturia Desmopressin 100–400mcg po/10–40mcg intranasally may help

Bowel problems

- Dysphagia Common. Fluids are more difficult to swallow than semisolids. Formal assessment by trained staff is essential. Feeding through nasogastric tube or percutaneous endoscopic gastrostomy (PEG) may be needed long or short term—in terminal disease (e.g. MND); weigh provision of nutrition against prolongation of poor-quality life
- Constipation Difficulty with defecation or bowel opening <2×/wk—↑ fluid intake and ↑ fibre in diet. If no improvement use po laxative ± regular suppositories/enemas
- Incontinence Exclude overflow due to constipation

Skin breakdown *Prevented by*: positioning; mobilization; good skin care; management of incontinence; pressure-relieving aids (e.g. special mattresses/cushions). Involve community nursing services.

Neurological assessment scales

A number of neurological assessment scales are in common use. Agreeing to use a formal, validated assessment scale enables comparison of observations between different team members, and also allows comparison of observations over time. Commonly used scales include:

Glasgow Coma Scale Assesses level of consciousness—➔ p. 1052

Motor scoring scale Assesses muscle power:

- 0—no muscle movement
- 1—muscle flicker but no movement
- 2—moves but not against gravity
- 3—supports limb against gravity but not resistance
- 4—able to overcome mild resistance (mild weakness)
- 5—able to overcome strong resistance (normal power)

Disability severity scales Assess the impact of a particular condition on the individual. These scales are usually condition specific, e.g. the Migraine Disability Assessment Scale (MIDAS) or Seizure Severity Questionnaire for patients with epilepsy. These scales are useful to gauge severity of symptoms and also monitor response to any treatments provided.

Daily living scales A number of scales are available that measure what the individual can do in practice. These may be:

- Non-disease specific e.g. Barthel Index, or
- Disease specific e.g. Oxford Stroke Handicap Scale

It is not really important which scale is used as long as everyone in the team uses the same scale for any given patient. Most use a graded Likert scale (e.g. 0–5) and rate activities such as:

- Mobility—walking, stairs, ability to transfer
- Personal care—dressing, washing
- Feeding—ability to prepare food, ability to feed self
- Toileting—ability to use the toilet, continence (bowels and bladder)

Quality of life scales Neurological conditions can have a profound impact on quality of life. Scales used to assess impact on quality of life may be completed by the patients themselves, or by the attending health professional. Examples include:

- Non-disease specific scales e.g. Euroqol EQ-5D
- Disease specific scales e.g. Quality of Life in Essential Tremor (QUEST); Quality of Life in Epilepsy (QUOLIE)

Cognitive function tests e.g. the General Practitioner Assessment of Cognition (GPCOG), or 6 Cognitive Impairment Test (6CIT—➔ p. 989).

Mental health scales e.g.:

- Anxiety GAD-2 (➔ p. 971)
- Depression NICE chronic disease depression screening questions (➔ p. 173); PHQ-9 (➔ p. 979)

Migraine disability assessment score (MIDAS) Used to assess the impact of migraine symptoms on lifestyle.

Instructions Please answer the following questions about ALL the headaches you have had over the last 3mo. If you did not do the activity in the last 3mo, write 0.

1. On how many days in the last 3mo did you miss work or school because of your headache?	☐ days
2. How many days in the last 3mo was your productivity at work or school ↓ by ≥½ because of your headaches? *(Do not include days you counted in question 1 where you missed work or school)*	☐ days
3. On how many days in the last 3mo did you not do household work[a] because of your headache?	☐ days
4. How many days in the last 3mo was your productivity in household work ↓ by ≥½ because of your headaches? *(Do not include days you counted in question 3 where you did not do household work)*	☐ days
5. On how many days in the last 3mo did you miss family, social, or leisure activities because of your headaches?	☐ days
MIDAS score TOTAL	☐ days
A. On how many days in the last 3mo did you have a headache? *(If a headache lasted more than 1 day, count each day)*	☐ days
B. On a scale of 0–10, on average how painful were these headaches? *(Where 0 = no pain at all, and 10 = pain as bad as can it be)*	☐

Questions A and B measure the frequency of the migraine and the severity of pain. They are not used to reach the MIDAS score, but provide extra information helpful for making treatment decisions.

Interpreting the MIDAS score

I	*Score: 0–5*	Minimal/infrequent disability	Tend to have little or no treatment needs. Can often manage with OTC medication. If infrequent severe attacks may require triptan
II	*Score: 6–10*	Mild/infrequent disability	May require medication for acute attacks, e.g. NSAID ± antiemetic or triptan
III	*Score: 11–20*	Moderate disability	Will need medication for acute attacks. Consider prophylaxis. Consider other causes for headaches, e.g. tension-type headache
IV	*Score: ≤21*	Severe disability	

[a] Unpaid work such as housework, shopping, and caring for children and others.

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Chapter 16

Dermatology

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Skin changes associated with internal conditions *614*

Skin assessment

History Use open questions at the start, becoming directive when necessary—clarify, reflect, facilitate, listen. *Ask about*:

Age and gender Influence probability of conditions (Table 16.1).

Presenting complaint Chronological account—when, where, and how the problem started. Ask directly about:

- Skin lesions What did the initial lesions look like? How have they evolved and extended?
- Associated symptoms Itching (➔ p. 566)? Sweating (➔ p. 575)? Systemic symptoms?
- Aggravating or relieving factors e.g. sunlight
- Past medical history Similar symptoms, atopy, systemic disease, e.g. rheumatoid arthritis, coeliac disease
- Family history Genetic skin problems, e.g. neurofibromatosis; other family members with similar symptoms, e.g. scabies
- Drug history New drugs (including OTC), immunosuppressants, drug allergies
- Occupation/hobbies Does the problem improve when away from work/hobbies? Could the problem have been caused as a result of work/hobbies?
- Previous treatments tried and result

Attitudes and beliefs How does the patient see the problem? What does he/she think is wrong? How does he/she think other people view the situation? What does the patient want you to do about it?

Examination Use a systematic approach to the skin lesions:

Distribution Figure 16.1

Individual lesion morphology A magnifying hand lens is often helpful in looking at individual lesions. Palpation is also important to determine consistency, depth, and texture. Skin lesions—➔ p. 564.

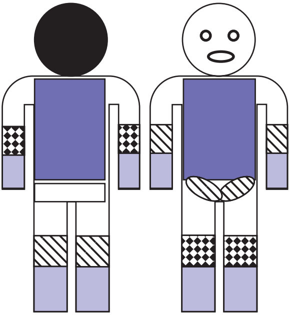
- Are lesions monomorphic (take one form—e.g. guttate psoriasis) or pleomorphic (take many forms—e.g. chickenpox)?
- Are there 2° changes on top of 1° lesions (e.g. excoriations)?
- How are lesions grouped locally (i.e. ring-shaped, linear, Koebner phenomenon)?

Check hair, nails, and mucous membranes Hair problems—➔ p. 574; nail changes—➔p. 576.

Consider general examination If examination of the skin suggests systemic cause.

Action

- Summarize the history back to the patient and give an opportunity for the patient to fill in any gaps
- Draw up a problem list and outline a management plan with the patient. Further investigations and interventions are guided by findings on history/examination—so a good history and examination are essential
- Set a review date



- Localized or generalized?
- Symmetrical? If so, are the lesions peripheral ▢ (e.g. lichen planus) or central ■ (e.g. pityriasis versicolor)?
- Do skin lesions involve the flexures ▧ (e.g. eczema) or extensor surfaces ▩ (e.g. psoriasis)?
- Are lesions limited to sun-exposed areas? ➲ p. 598
- Are lesions linear or ring-shaped? ➲ p. 564
- Is the distribution dermatomal, e.g. shingles?
- Does the problem affect only one region, e.g. axilla, face, groin, foot?

Figure 16.1 Assessing distribution of skin lesions

Table 16.1 Age, gender, and probability of different skin conditions

	Skin conditions that are more common
Male	Seborrhoeic dermatitis; dermatitis herpetiformis; porphyria cutanea tarda; polyarteritis nodosa; pruritus ani; tinea pedis and cruris; mycosis fungoides; squamous cell carcinoma
Female	Palmoplantar pustulosis; lichen sclerosus; lupus erythematosus; systemic sclerosis; morphoea; rosacea; dermatitis artefacta; venous ulceration; malignant melanoma
Child	Port wine stain; strawberry naevus; ichthyosis; erythropoietic porphyria; epidermolysis bullosa; atopic eczema; infantile seborrhoeic dermatitis; urticaria pigmentosa; viral infection, e.g. chickenpox, warts, molluscum contagiosum; head lice; impetigo
Adolescent	Melanocytic naevi; acne; psoriasis (particularly guttate); seborrhoeic dermatitis; pityriasis rosea; vitiligo
Early adult	Psoriasis; seborrhoeic dermatitis; lichen planus; dermatitis herpetiformis; lupus erythematosus; vitiligo; pityriasis versicolor
Middle age	Lichen planus; rosacea; pemphigus vulgaris; venous ulceration; malignant melanoma; basal cell carcinoma; mycosis fungoides; porphyria cutanea tarda
Old age	Asteatotic eczema; senile pruritus; bullous pemphigoid; venous and arterial ulcers; seborrhoeic warts; solar elastosis and keratosis; Campbell de Morgan spots; basal cell and squamous cell carcinomas; herpes zoster

Treatment of skin conditions

Skin conditions are usually treated with topical creams and lotions. Consider the vehicle as well as the active ingredient. In primary care the choice is usually between creams or ointments.

- **Creams** Emulsions of oil and water. Well absorbed into the skin, less greasy and easier to apply than ointments
- **Ointments** Greasy preparations suitable for chronic, dry lesions

Alternatives include: applications, mousses, foams, gels, lotions, and pastes.

Emollients e.g. white soft paraffin. Useful for all dry or scaling disorders to soothe, smooth, and hydrate the skin—apply in the direction of hair growth. Effects are short-lived—advise frequent application even after improvement occurs. Quantities to prescribe—Table 16.2.

- Severity of the condition, patient preference, and site of application guide choice of emollient. ❗ Some ingredients may cause sensitization (Box 16.1)—suspect if an eczematous reaction occurs
- Preparations such as aqueous cream and emulsifying ointment can be used as soap substitutes for hand washing and in the bath. Addition of an emollient bath oil (e.g. Oilatum® or Balneum®) may also be helpful. ❗ Aqueous cream should not be used as an emollient
- Avoid preparations containing an antibacterial (e.g. Dermol®) unless infection is present or a frequent complication
- Using a preparation with added urea (e.g. Balneum® Plus, Eucerin®) may improve hydration for scaling conditions or in elderly patients

Topical corticosteroids

- Used to suppress inflammatory conditions of the skin, e.g. eczema, when other measures, such as emollients, are ineffective used alone. Use the least potent preparation that is effective (Table 16.3). Apply a thin layer just to affected areas >30min before or after emollients once daily. Quantities to prescribe—Table 16.2
- Creams are suitable for moist or weeping lesions and ointments for dry, lichenified, or scaly lesions or where a more occlusive effect is wanted. Lotions may be useful when minimal application to a large or hair-bearing area is needed or for the treatment of exudative lesions
- Inclusion of urea or salicylic acid ↑ penetration of the corticosteroid

Cautions and contraindications Topical steroids:

- Are of no value in the treatment of urticaria
- Are contraindicated for rosacea and not recommended for acne
- May worsen ulcerated or secondarily infected lesions
- Should not be used indiscriminately for pruritus—they will only be of benefit if inflammation is causing the itch
- Should not be used long-term (>7–14d) for children or on the face (and keep away from eyes)

❗ For perioral inflammatory lesions use hydrocortisone 1% for ≤7d or, if infected (e.g. angular cheilitis), hydrocortisone + miconazole cream.

⚠ Potent topical or systemic steroids used to treat patients with psoriasis can result in rebound relapse, development of generalized pustular psoriasis, and/or local and systemic toxicity.

Table 16.2 Quantities of emollients and corticosteroids to prescribe

Area affected	Emollient[a]		Topical steroid (g)[b]
	Cream/ointment (g)	Lotion (mL)	
Face/neck	10–30	100	15–30
Both hands	25–50	200	15–30
Scalp	50–100	200	15–30
Both arms	100–200	200	30–60
Both legs	100–200	200	100
Trunk	400	500	100
Groins/genitalia	15–25	100	15–30

[a] Amounts are for an adult for 2×/d application for 1wk.

[b] Amounts are for an adult for once-daily application for 2wk.

Box 16.1 Ingredients that may cause skin sensitization

Benzyl alcohol
Butylated hydroxyanisole
Butylated hydroxytoluene
Cetostearyl alcohol (including acetyl and stearyl alcohol)
Chlorocresol
Edetic acid (EDTA)
Ethylenediamine
Fragrances
Hydroxybenzoates (parabens)
Imidurea
Isopropyl palmitate
Methylchloroisothiazolinone (MCI)
Methylisothiazolinone (MI)
N-(3-chloroallyl) hexaminium chloride
Polysorbates
Propylene glycol
Sodium metabisulphite
Sorbic acid
Wool fat/related substances, including lanolin

Table 16.3 Topical corticosteroid preparation potencies

Potency	Examples
Mild	Hydrocortisone 0.1–2.5%, Dioderm®, Mildison®, Synalar® 1 in 10 • *With antimicrobials* Canesten® HC, Fucidin® H, Timodine® • *With crotamiton* Eurax-Hydrocortisone®
Moderate	Betamethasone valerate 0.025% (Betnovate®-RD), Eumovate® • *With antimicrobials* Trimovate® • *With urea* Alphaderm®
Potent	Betamethasone valerate 0.1% (Betnovate®), Betacap®, Elocon®, Locoid®, Synalar® • *With antimicrobials* Aureocort®, Betnovate®-C or -N, Fucibet® • *With salicylic acid* Diprosalic®
Very potent	Dermovate®, Nerisone® Forte • *With antimicrobials* Dermovate®-N

❗ 1 fingertip unit (distance from the tip of the adult index finger to the 1st crease) of steroid cream is sufficient to cover an area 2× the size of the flat adult palm.

Changes in skin colour and eruptions

Pallor Non-specific sign which may be racial, familial, or cosmetic. Pathology suggested includes anaemia, shock, Stokes–Adams attack, vasovagal faint, myxoedema, hypopituitarism, and albinism.

Erythema ➔ p. 568 **Hypo- and hyperpigmentation** ➔ p. 572

Linear lesions *Consider*:
- Koebner phenomenon (lesions arise in area of injury, e.g. in scratches)—occurs in psoriasis, vitiligo, lichen planus
- Symptomatic dermographism
- Self-inflicted trauma—dermatitis artefacta
- Reaction to garden plants—psoralen-induced phytophotodermatitis
- Impetigo—may spread along scratch marks
- Herpes zoster—at the edge of a dermatome

Ring-shaped (annular) lesions *Consider*:
- Psoriasis
- Discoid eczema
- Urticaria
- Pityriasis rosea
- Lichen planus
- Orf
- Granuloma annulare
- Erythema multiforme
- Basal cell carcinoma
- Fungal infection, e.g. ringworm
- Erythema migrans associated with Lyme disease
- Burns (especially on a child—may be non-accidental injury)
- Cutaneous T-cell lymphoma (rare)

White patches Consider all causes of patchy hypopigmentation:
- Vitiligo
- After inflammation—cryotherapy, eczema, psoriasis, morphoea
- Pityriasis alba—white post-inflammatory patch on a child's face. No treatment needed
- Exposure to some chemicals—substituted phenols, hydroquinone
- Certain infections—pityriasis versicolor
- Tuberous sclerosis—'Ash leaf' patches
- Halo naevus—pale area around a mole
- Piebaldism—from birth—associated with a white forelock
- Extensive hyperpigmentation, e.g. chloasma—patches of normal skin may appear hypopigmented

White spots *Consider*:
- Pustules/whiteheads, e.g. due to acne, folliculitis, or rosacea
- Molluscum contagiosum—white spots with a pearl-like appearance
- Milia (➔ p. 595)—small white spots usually on upper arms/face of children—resolve spontaneously

Brown spots *Consider*:
- Freckles
- Moles
- Lentigos—like freckles but darker and not affected by sunlight
- Melanoma
- Café au lait spots—>5 associated with neurofibromatosis
- Basal cell carcinoma
- Seborrhoeic warts
- Dermatofibroma
- Systemic disease
- Addison's disease
- Acanthosis nigrans
- Haemochromatosis

Scaling

- Silvery scaling on the surface of red patches Psoriasis
- Fine scaling accompanied with rash Pityriasis; 2° syphilis
- Coarse, scaly skin with no rash Ichthyosis
- Localized, not itchy Bowen's disease

Yellow crusting Usually due to staphylococcal infection (impetigo).

Telangiectasia Dilated distal venule/arteriole (spider naevus). *Causes*:

- Congenital e.g. hereditary haemorrhagic telangiectasia
- Venous disease in the leg e.g. venous stars
- Rosacea Facial
- Excess oestrogen e.g. liver disease; CHC; pregnancy
- Skin atrophy e.g. ageing skin, radiation dermatitis, topical steroids
- Surface of BCC

Spider naevi Small, red lesions (barely visible <0.5cm diameter) in superior vena cava distribution, i.e. on the arms, neck, and chest wall. Large arteriole with numerous small vessels radiating from it giving the appearance of a spider—hence the name. Pressure applied to the central arteriole (e.g. with a pointed object) causes blanching of the whole lesion. >2 spider naevi is abnormal. *Causes*:

- Cirrhosis—most frequently, alcoholic
- Oestrogen excess—chronic liver disease or medication
- Rheumatoid arthritis—rarely
- Viral hepatitis—transient
- Pregnancy—usually disappear in the final trimester

Blisters ➔ p. 566

Subcutaneous nodules *Consider*:

- Cysts
- Rheumatoid arthritis
- Xanthelasma
- Tumour
- Neurofibroma
- Granuloma annulare
- Furuncle
- Sarcoid
- Polyarteritis

Purpura Blue-brown discolouration of the skin due to bleeding within it. Petechiae are small dot-like purpura while ecchymoses are more extensive. Treat the cause:

- Idiopathic e.g. pigmented purpuric dermatosis (brownish punctate lesions on the legs)
- Vessel wall defects Vasculitis, paraproteinaemia, infection (e.g. meningococcal meningitis, septicaemia, glandular fever), ↑ intravascular pressure (e.g. venous disease)
- Clotting defects Abnormal platelet function; thrombocytopenia; anticoagulant therapy; coagulation factor deficiency
- Defective dermal support Dermal atrophy (e.g. ageing, steroids, disease); scurvy (vitamin C deficiency)

⚠ *Referral of patients with purpura*

- Admit unwell patients with new purpura/petechiae as an emergency
- Refer well children/young adults with unexplained petechiae immediately to be seen the same day[N]
- For well, older adults with unexplained bruising, bleeding, or purpura, check FBC, blood film, clotting screen, and ESR/viscosity/CRP[N]

Itching and blistering of the skin

Itching (pruritus) In any patient presenting with pruritus or itch, ask: *are there skin lesions present?*

Skin lesions present Search for unexcoriated lesions. Investigations are not usually needed. Exceptions are patch testing for contact dermatitis and skin biopsy for dermatitis herpetiformis. *Causes*:

- Urticaria
- Contact dermatitis and allergies to food and drugs
- Polymorphic light reaction
- Infections—viral e.g. chickenpox; fungal
- Dermatitis herpetiformis
- Lichen planus
- Senile atrophy
- Skin infestations, e.g. scabies, pediculosis, insect bites
- Psychological causes (excessive scratching/rubbing causes lichenification of the skin)

Skin lesions absent Large differential diagnosis. Look for pallor, jaundice, weight ↓, LN enlargement, and abdominal organomegaly. Investigate as necessary—consider urinalysis (dipstick/MSU), FBC, ESR/CRP, serum ferritin, LFTs, bone profile (including alk phos and Ca^{2+}), U&E, Cr and eGFR, HbA1c, TFTs, myeloma screen, and CXR. Consider HIV and hepatitis screen. If still undiagnosed—refer. *Causes*:

- Hepatic—obstructive jaundice, pregnancy
- Endocrine—DM, thyrotoxicosis, hypothyroidism, hyperparathyroidism
- Renal—chronic renal failure
- Haematological—polycythaemia vera, iron deficiency, leukaemia, Hodgkin's disease, myeloma
- Malignancy—any carcinoma
- Drug allergies
- Psychological—obsessive states, schizophrenia
- Rare causes—HIV, diabetes insipidus, roundworm infection

Blisters Result from separation of skin layers. Type of blister depends on level of cleavage of the skin—subcorneal or intraepidermal blisters rupture easily, subepidermal blisters are much tougher. *Causes*:

- **Subcorneal** Pustular psoriasis (➜ p. 590), bullous impetigo (➜ p. 606)
- **Intraepidermal** Eczema (➜ p. 578), HSV (➜ p. 608), varicella zoster virus—chickenpox (➜ p. 629) or shingles (➜ p. 628), pemphigus, friction
- **Subepidermal** Cold or heat injury (burns—➜ p. 1096, pemphigoid, dermatitis herpetiformis, linear IgA disease
- **Other** Insect bites (may cause cleavage at any level)

Pemphigoid[G] Autoimmune disorder.

- **Bullous pemphigoid** Usually elderly. An urticated rash may precede onset of blistering. Large, tense blisters arise on red/normal skin on the limbs, trunk and flexures. Oral lesions in 20–30%. May be restricted to one site, e.g. lower leg. *Differential diagnosis*: pemphigus, dermatitis herpetiformis, linear IgA disease, epidermolysis bullosa acquisita
- **Cicatricial pemphigoid** Mainly affects mucous membranes in the eyes/mouth. Scarring results in visual loss. Refer to ophthalmology
- **Pemphigoid gestationis** Rare but characteristic bullous eruption associated with pregnancy. Remits after delivery but often recurs in subsequent pregnancies. ➜ p. 791

Management Check blood for skin antibodies—pemphigoid (skin basement membrane) antibody—present in 75% with bullous pemphigoid. Refer to dermatology for skin biopsy and confirmation of diagnosis. Treatment is usually with oral steroids (prednisolone 30–60mg daily initially—reducing as symptoms improve). Other treatments include antibiotics and nicotinamide, azathioprine, or other immunosuppressants.

Prognosis Self-limiting in 50%—steroids are often stopped after ~2y.

Pemphigus^{G} Uncommon, autoimmune disorder affecting skin and mucous membranes. Affects adults (peak incidence 30–70y). *Cause*: 90% have detectable circulating autoantibodies. Associated with other autoimmune disorders, e.g. myasthenia gravis.

Presentation 50% present with oral lesions. Suspect in anyone presenting with mucocutaneous erosions/blisters. Flaccid superficial blisters then appear—sometimes months later—over scalp, face, back, chest, and flexures. As blisters are fragile they burst early and the condition may present as crusted erosions. Untreated the condition is progressive.

Management Check blood for skin antibodies—pemphigus (skin desmosome) antibodies are present in >80%. Refer to dermatology. Treatment is with high-dose systemic steroids or other immunosuppressive agents. Treatment is continued long term although occasional remissions occur.

Dermatitis herpetiformis ♂ > ♀ (2:1). *Peak incidence*: 3rd/4th decade. Consists of itchy vesicular skin rash on elbows (extensor surface), knees, buttocks, and scalp which are often broken by scratching to leave excoriations. Closely related to coeliac disease (➔ p. 382); up to 20% of patients with coeliac disease have dermatitis herpetiformis, but classic symptoms of coeliac disease are uncommon. *Differential diagnosis*: scabies, eczema, linear IgA disease.

Management Check IgA levels and TTG/EMA. Refer to dermatology for skin biopsy to confirm diagnosis. Responds to withdrawal of gluten—although may take up to 1y. Controlled in the interim with dapsone or sulfapyridine.

Epidermolysis bullosa A group of genetically inherited diseases characterized by blistering on minimal trauma. Range from being mild and trivial to being incompatible with life. The most common form is *simple epidermolysis bullosa* (autosomal dominant)—blistering is caused by friction, is mild and limited to hands and feet. Patients are advised to avoid trauma.

Linear IgA disease Rare condition of blisters and urticarial lesions on the back and extensor surfaces. Refer to dermatology. Responds to potent topical steroids (if mild), erythromycin, or dapsone.

Staphylococcal scalded skin syndrome ➔ p. 883

Further information

Primary Care Dermatology Society (2016) Blistering (bullous) disorders—an overview. 🖱 www.pcds.org.uk/clinical-guidance/bullous-disorders-an-overview

Erythema

Erythema is redness of the skin—usually due to vasodilation. It may be localized (e.g. pregnancy—on the palms), generalized (e.g. flushing), or take the form of a red rash (e.g. drug eruption, viral exanthem).

⚠ **Erythroderma (exfoliative dermatitis)** Erythema affecting >90% skin surface. Rare, but systemic effects of skin failure are potentially fatal. ♂:♀ ≈2:1. Typical patient is middle-aged or elderly. Patchy erythema becomes universal in <48h. Accompanied by fever, shivering, and malaise. 2–6d later scaling appears. The skin is hot, red, itchy, dry, thickened, and feels tight. There may be oedema/oozing. Hair and nails may be shed. Admit as an acute medical emergency.

Causes Eczema (40%); psoriasis (25%); lymphoma (15%); drug eruption (10%); other skin disease (2%); unknown (8%).

Staphylococcal scalded skin syndrome ➔ p. 883

Erysipelas and cellulitis ➔ p. 606

Flushing Generalized erythema due to vasodilation. Common and usually benign. Tends to affect face, neck, and upper trunk. *Cause*:

- Physiological e.g. exertion, heat
- Emotion e.g. anger, anxiety, embarrassment
- Foods e.g. spices, chillies, alcohol
- Endocrine e.g. menopause, Cushing's syndrome
- Drugs e.g. opioids, tamoxifen, danazol, GnRH analogues, clomifene, nitrates, calcium-channel blockers
- Dermatological Rosacea (unknown mechanism); contact dermatitis
- Inflammatory SLE; dermatomyositis
- Infection e.g. slapped cheek syndrome (fifth disease); cellulitis/erysipelas
- Tumour Pancreatic tumours, medullary thyroid cancer, carcinoid, phaeochromocytoma

Management Treat cause if possible (e.g. avoid alcohol, HRT). Embarrassing flushing may be helped with propranolol (e.g. 40mg od/bd) or clonidine (e.g. 50 micrograms bd). If severe and disabling and no response to conservative measures, consider referral to dermatology.

Palmar erythema Generalized reddening of the palms associated with pregnancy, liver disease, and polycythaemia.

Erythema nodosum Tender erythematous nodules 1–5cm diameter on extensor surfaces of limbs—especially shins (Figure 16.2) ± ankle and wrist arthritis ± fever. ♀:♂ ≈3:1. Resolves in <8wk; non-scarring. No treatment needed; analgesia and mild compression may ease symptoms.

Associations 20% of cases are idiopathic with no associations.

- Streptococcal infection
- Drugs, e.g. oral contraceptives, sulfonamides
- Acute sarcoidosis
- Inflammatory bowel disease—UC, Crohn's
- Malignancy
- TB

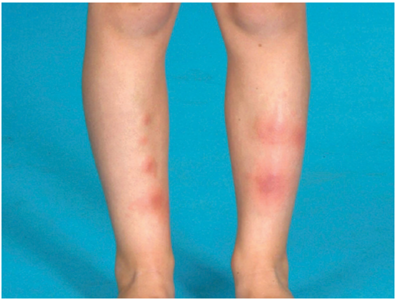


Figure 16.2 Erythematous nodules on the shins of a patient with erythema nodosum

Reproduced from Foster H. et al., *Paediatric Rheumatology* (2012) with permission from Oxford University Press

Erythema multiforme Immune-mediated disease characterized by target lesions on hands and feet (Figure 16.3). *Causes*:

- Idiopathic (50%)
- Infective Streptococcal, HSV, hepatitis B, mycoplasma
- Drugs Penicillin, sulfonamide, barbiturate
- Other SLE, pregnancy, malignancy

Presentation Target lesions (red rings with central pale or purple area) on hands and feet. New lesions appear for 2–3wk. Frequently oral, conjunctival, and genital mucosa is affected—if severe, termed *Stevens–Johnson syndrome*.

Differential diagnosis Toxic erythema, toxic epidermal necrolysis, Sweet's disease, urticaria, pemphigoid.

Management Identification and removal of the underlying cause. Mild cases resolve spontaneously and require symptomatic measures only. Admit if extensive involvement.

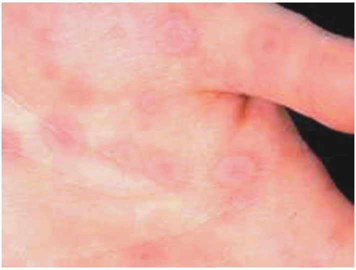


Figure 16.3 Typical target lesions of erythema multiforme

Reproduced with permission from Sladden MJ, *et al*. Common skin infections in children: Folliculitis and herpes. *BMJ*, **13**:265–308. Copyright © 2005 BMJ Publishing Group Limited. All rights reserved. https://www.bmj.com/content/331/Suppl_S1/0507274.full

Rosacea Relapsing–remitting chronic inflammatory facial dermatosis characterized by erythema and pustules. Most common in middle age (30–50y) and in fair-skinned people of Northern European descent. ♀ > ♂ (≈3:2). No cure. *Cause*: unknown—although possible associations with the face mite *Demodex folliculorum, Helicobacter pylori* infection, and migraine headaches.

Presentation Earliest symptom is flushing. Erythema, telangiectasia, papules, pustules ± lymphoedema affect cheeks, nose, forehead, and chin (Figure 16.4a). No comedones.

Aggravating factors Sun exposure (61%); emotional stress (60%); hot weather (53%); alcohol (45%); spicy foods (43%); exercise (39%); cold weather or wind (38/36%); hot baths (37%); hot drinks (36%); cosmetics/skin care products (24%); topical steroids.

Complications Rhinophyma (bulbous appearance of nose—Figure 16.4b); eye involvement—blepharitis, dry eye, and conjunctivitis.

Differential diagnosis Acne (lacks comedones and older age group); contact dermatitis; SLE; photosensitive eruptions; seborrhoeic dermatitis.

Management
- Avoid triggers
- Antibiotics—repeated treatment is usually needed over many years with prolonged courses of topical or systemic antibiotics (e.g. topical ivermectin od for 3mo; topical metronidazole 0.75% gel/cream bd for 3mo; lymecycline 408mg od po). Rebound may occur if antibiotics are stopped suddenly
- Refer to dermatology if rhinophyma, ocular complications, or failure to respond to treatment in general practice

Livedo reticularis Marbled, patterned cyanosis of the skin. If not reversible by warming, investigate and treat the cause. *Causes*: physiological (e.g. cold); vasculitis (e.g. SLE); hyperviscosity.

Chilblains Inflamed and painful purple pink swellings on fingers, toes, or ears. Appear in response to cold. ♀ > ♂. Advise warm housing/clothing, gloves, and woolly socks. In severe cases oral nifedipine may help.

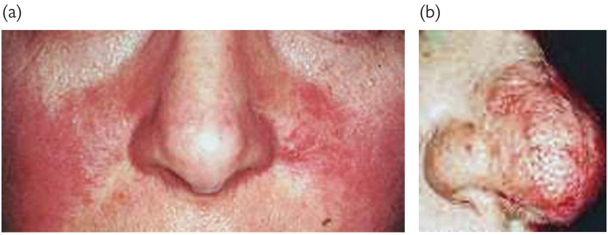


Figure 16.4 Rosacea: (a) redness and pustules on nose, cheeks and forehead and (b) rhinophyma

Reproduced with permission of the National Rosacea Society from Blount BW, *et al*. Rosacea: a common, yet commonly overlooked, condition. *Am Fam Physician*, 2002;**66**(3):435–40.

Erythema ab igne Reticulate pigmented erythema due to heat-induced damage. Common in the elderly—especially from sitting in front of the fire or using hot water bottles to alleviate pain. In younger patients may be caused by laptop computers balanced on thighs. Explain the cause. Resolves spontaneously.

Viral rash Common, particularly in children. Appears suddenly (over hours) and is associated with symptoms of the underlying viral infection. The rash may take many forms but is usually widespread, red, maculopapular and blanches on pressure. In most cases the underlying virus cannot be identified, but viral infections with characteristic rashes include:

- Chickenpox ➔ p. 629
- Measles ➔ p. 629
- Rubella ➔ p. 629
- Fifth disease (slapped cheek) ➔ p. 629
- Roseola infantum ➔ p. 629
- Hand, foot, and mouth ➔ p. 629

Management In all cases, no specific treatment is needed for the rash—treat the underlying viral infection symptomatically.

Rash illness in pregnancy ➔ p. 790

Lyme disease *Cause: Borrelia burgdorferi. Spread*: transmitted by ticks—usually from deer or sheep. Presents with:

- Erythema migrans (75%—Figure 16.5)—a red macule/papule on the upper arm, leg, or trunk 7–10d after a tick bite which expands over days/weeks to form a ring with central clearing; diameter can be up to 50cm; further smaller lesions then develop elsewhere
- Flu-like illness ± arthralgia
- Lymphadenopathy ± splenomegaly

Symptoms are typically intermittent and changing. Complications include neurological abnormalities, aseptic meningitis, myocarditis, and arthritis.

Management If symptoms are present, start antibiotics and confirm diagnosis with serology (❗ false –ve results are common). Treatment is usually with 2–3wk course of doxycycline—take microbiology advice. 💣 Treatment with antibiotics after a tick bite but before symptoms have arisen is controversial.

Removal of ticks ➔ p. 1091

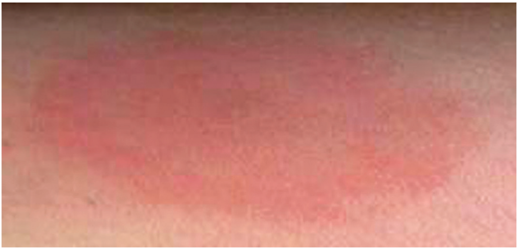


Figure 16.5 Erythema migrans following a tick bite

Pigmentation disorders

Hypopigmentation Lack of skin pigmentation. May be:

- **Generalized** Albinism; phenylketonuria; hypopituitarism
- **Patchy** Vitiligo; tuberous sclerosis; morphoea; pityriasis alba; or after inflammation (e.g. post-cryotherapy, 2° to eczema or psoriasis), infection (e.g. pityriasis versicolor), or exposure to chemicals (substituted phenols, hydroquinone)
- **Around a mole** Halo naevus

Hyperpigmentation Excess skin pigmentation. May be:

- **Genetic** Racial; freckles; neurofibromatosis; Peutz–Jeghers syndrome
- **Due to drugs** Amiodarone (blue-grey pigmentation of sun-exposed areas); psoralens; minocycline (blue-black pigmentation in scars and buccal mucosa); chloroquine (blue-grey pigmentation of face and arms); chlorpromazine (grey pigment in sun-exposed sites); cytotoxics
- **Endocrine** Addison's disease; chloasma; Cushing's syndrome
- **Nutritional** Ingestion of carrots (carotenaemia); malabsorption
- **Post-inflammatory** Varicose eczema; lichen planus; systemic sclerosis
- **Other** Benign naevi; malignant melanoma; chronic renal failure (lemon yellow); liver disease (jaundice); acanthosis nigricans

Freckles and lentigines

- **Freckles** Small, light-brown macules—typically facial—which darken in the sun. They are common, particularly in people with red hair, and develop in childhood. Require no treatment
- **Lentigines** Also brown macules but more scattered and do not darken in the sun. Most common in elderly sun-exposed skin

Chloasma Patterned macular symmetrical facial pigmentation, usually involving the forehead and/or cheeks (Figure 16.6), ♀ >> ♂. Peak age 20–40y. Genetic predisposition. Affects dark skins > fair skins.

Risk factors UV radiation; pregnancy (usually fades after delivery); taking CHC or depot contraceptives (may be slow to fade if stopped); use of cosmetics, perfumes, or deodorant soap (phototoxic reaction).

Management Reassurance is normally all that is needed. *Consider*:

- Stopping hormonal contraception
- Avoid irritating the skin with strong soaps/abrasive cleaners
- Vigorous photoprotection using sunscreen and hat
- 20% azelaic acid cream or 0.05% tretinoin cream may be used but requires application for >6mo to have any effect
- Camouflage cosmetics and/or fake tanning products
- Specialist treatments—chemical peels, laser resurfacing, Pigmanorm® cream—refer for expert advice

Albinism *Prevalence*: 1 in 20,000. Rare genetic syndrome (autosomal recessive inheritance) in which the melanocytes are unable to produce skin, hair, or eye pigment. Patients have white hair, pale skin, pink eyes, poor sight, photophobia, and nystagmus. Several different varieties exist.

Management Strict sun avoidance, sunglasses, sunscreens, refer any skin lesions for biopsy (↑ risk squamous cell carcinoma).

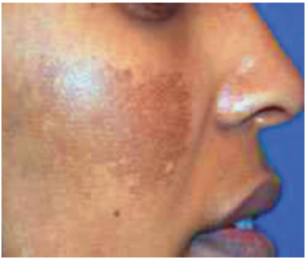


Figure 16.6 Chloasma of the cheek

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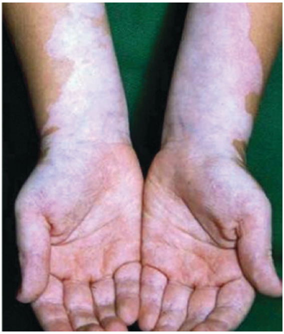

Figure 16.7 Vitiligo on the forearms

Vitiligo Affects 1% of the population. ♂ = ♀. *Peak age of onset*: 10–30y. *Cause*: autoimmune; 18% have a family history. *Associations*: DM, alopecia areata, pernicious anaemia, Addison's disease, and thyroid disease.

Presentation May be precipitated by injury or sunburn. Presents as smooth, sharply defined white macules or patches which contain no melanocytes (Figure 16.7). Skin appears bright white under Wood's light (if available). Often symmetrical distribution. Hair may be affected. *Most common sites*: hands, wrists, knees, neck, face around eyes and mouth.

Differential diagnosis Post-inflammatory hypopigmentation, piebaldism, pityriasis alba, chemical exposure, leprosy.

Management Prognosis is variable; spontaneous remission is unusual; often pigment loss is progressive. Advise use of high-factor sunscreens for affected areas (ACBS prescription); camouflage cosmetics (refer to Changing Faces for advice on application, ACBS prescription). Consider referral to dermatology for consideration of topical steroid, topical calcineurin inhibitors (tacrolimus, pimecrolimus—unlicensed), or narrowband UVB (TLO1) phototherapy (helps 67% but relapse may occur after treatment ends).

Morphoea (localized scleroderma) ♀:♂ ≈3:1. Cutaneous localized form of scleroderma. Pathologically distinct from lesions of systemic sclerosis. Internal disease is not associated. *Cause*: autoimmune but not fully understood—may follow trauma. Presents with round/oval plaques of induration and erythema, which become smooth, shiny, and white, with violet borders. Eventually leaves atrophic hairless pigmented patches. Affects trunk/proximal limbs. No established treatment—usually resolves spontaneously in 3–5y; potent topical steroids may be tried.

Further information

Primary Care Dermatology Society Hypo- and hyperpigmentation. 🖱 www.pcds.org.uk/p/general-dermatology-appearance#1

Patient support

Changing Faces Skin Camouflage Service 🖱 www.changingfaces.org.uk

Hair and sweat gland problems

Hair loss or alopecia Treat according to cause. *Differential diagnosis*:

- **Diffuse non-scarring** ♂/♀ pattern baldness; telogen effluvium
- **Localized non-scarring** Alopecia areata; ringworm (kerion may scar); traumatic; hair pulling; traction; SLE; 2° syphilis
- **Scarring** Burns; radiation; shingles; tertiary syphilis; lupus erythematosus; morphoea; lichen planus; kerion; folliculitis decalvans

Telogen effluvium

- **Acute** Occurs ~3mo after a trigger, e.g. pregnancy, fever, surgery, weight ↓ (crash diet), stress—if the trigger is not repeated, hair regrows in 3–6mo
- **Chronic** Occurs in middle-aged ♀. Cause unknown. Large amounts of hair shedding but appearance of a 'normal head of hair'. Reassure
- **Secondary** Check FBC, ferritin, TFTs. Cause may be endocrine (hypothyroidism, hypopituitarism, hypoadrenalism); drug induced (e.g. chemotherapy); or nutritional (iron deficiency—treat if ferritin <40 micrograms/L; zinc deficiency)

Pattern baldness Affects ♂ at any age (androgenic alopecia) and usually postmenopausal ♀. Strong genetic predisposition. For ♀, if evidence of virilization (e.g. acne, hirsutism), further investigation is needed. *Treatment options*:

- No treatment
- Wig
- Hair products containing nanofibers to make hair appear thicker
- Topical minoxidil—thickens/↑ density of remaining hair. Use 5% foam/solution for ♂; 2% solution for ♀. Give a 6–12mo trial. Helps ~40%
- Finasteride (♂)—1mg od or 5mg/wk. Reassess after ~6mo. Helps ~two-thirds
- Spironolactone (♀)—50–200mg daily ± minoxidil ↓ hair loss in ~75%
- Surgery—follicular unit transplantation. Variable outcomes

❗ Drug treatment is not available on NHS prescription; effect is lost if treatment is discontinued. Surgery is not available through the NHS.

Alopecia areata[G] Common autoimmune disease affecting hair follicles ± nails (~10%—causes pitting). 20% have a family history. Patches of hair loss usually on the scalp but can affect any hair-bearing skin. Short broken (exclamation mark) hairs are seen at the margins of active areas of alopecia; yellow dots may be seen around hair follicles. Consider alternative diagnosis of tinea capitis if scales/erythema present.

Management Investigation is usually unnecessary. If mild hair loss, reassure and monitor hair loss; if more severe, refer to dermatology. Specialist treatment options include topical/locally injected/systemic steroids ± contact immunotherapy. ~40% recover in <1y; 20% lose all scalp hair—recovery in these cases is unusual (<10%). Psychological support may be needed.

Hirsutism Excess hair in androgenic distribution—➔ p. 312.

Hypertrichosis Excess hair in non-androgenic distribution—usually face and trunk, although can be generalized. Mostly drug induced (e.g. phenytoin, ciclosporin, minoxidil). If not, investigate to find other causes: malnutrition, anorexia nervosa, porphyria cutanea tarda, malignancy. If a cause cannot be found, treat symptomatically with electrolysis, bleaching, waxing, ± depilatories.

Local hypertrichosis Can be associated with topical steroid usage, be over a melanocytic naevus or associated with spina bifida occulta.

Hidradenitis suppurativa Chronic, autoimmune, inflammatory condition of sweat glands in axilla, groin, and perineum. ♀:♂ ≈3:1. May be a family history. Nodules, abscesses, cysts, and sinuses form → scarring. Treat with topical antiseptics (e.g. 4% chlorhexidine wash) or antiseptic emollients (e.g. Dermol 500® lotion), systemic antibiotics (e.g. lymecycline 408mg od), ± surgical drainage/excision. Refer to dermatology if these measures fail.

Hyperhidrosis Sweating, or perspiration, is normal and essential for temperature control. The amount people sweat varies enormously. Usually sweating can be controlled with shop-bought antiperspirants and is only excessive when it cannot be controlled and interferes with the patient's quality of life. Excessive sweating may be focal or generalized.

Generalized hyperhidrosis Most likely to occur 2° to other medical conditions. Where possible treat the cause:

- **Physiological** After and during exercise; hot, humid conditions; emotional response (e.g. anxiety)
- **Infection** Can occur with any bacterial or viral infection. Consider malaria if recent history of travel
- **Non-infective** Menopause; Parkinson's disease; DM; thyrotoxicosis; phaeochromocytoma; lymphoma; leukaemia; drug/drug withdrawal

If no cause is found, consider prescribing propantheline 15mg od–30mg tds. *Alternatives (all off licence) include*: oxybutynin MR 10mg/d; propranolol 40mg tds; or diltiazem 60mg tds. ❗ May take 6wk to work.

Focal hyperhidrosis Affects ~5% of the population. Usually a primary condition. Mainly affects axillae, palms, soles of feet, and/or face. Onset is typically in the teenage years. Distressing and socially disabling.

- Advice for patients—avoid clothing made of elastane, nylon, and other man-made fibres and tight clothing; wear colours that do not show the sweat (e.g. white, black); use emollient washes/moisturizers rather than soap; identify trigger factors for sweating (e.g. alcohol, crowded rooms) and avoid those situations. Specialized products may be helpful (🖱 www.sweathelp.co.uk)
- Treat topically with 20% aluminium chloride (e.g. Anhydrol forte®). Apply to clean skin at night—wash off in morning. ↓ frequency of application as symptoms subside. Treat local irritation with topical steroid, e.g. hydrocortisone 1%. Absorbent dusting powder may help axillary/plantar sweating
- Consider a trial of drug therapy as for generalized hyperhidrosis
- Refer to dermatology if not responding

Specialist treatment Iontophoresis; botulinum toxin injections; endoscopic transthoracic sympathectomy for severe palmar hyperhidrosis.

Further information

Primary Care Dermatology Society

- Alopecia (2019). 🖱 www.pcds.org.uk/clinical-guidance/alopecia-an-overview
- Hirsutism (2017). 🖱 www.pcds.org.uk/clinical-guidance/hirsutism
- Hyperhidrosis (excess sweating) (2017). 🖱 www.pcds.org.uk/clinical-guidance/hyperhidrosis

Nail changes

Nail changes may be due to nail disease or indicate other dermatological or systemic disease (Table 16.4).

Assessment

- **Take a history** Duration, initial changes, evolution of changes, other systemic or local symptoms, family history, drug and alcohol history, occupation, hobbies
- **Examine** Colour, shape, extent and pattern of involvement; consider examination/investigation for other skin or systemic disease (guided by history and appearance of nails)

Table 16.4 Nail changes: descriptions and causes

Change	Description of nail	Differential diagnosis	
Colour	Black transverse bands	Cytotoxic drugs	
	Blue	Cyanosis Antimalarials Haematoma	Phenothiazines Wilson's disease
	Blue-green	*Pseudomonas* infection	
	Brown	Fungal infection Cigarette staining Chlorpromazine	Gold Addison's disease
	Brown 'oil stain' patches	Psoriasis	
	Brown longitudinal streak	Melanocytic naevus Malignant melanoma Addison's disease	
	Red streaks—'splinter haemorrhages'	Infective endocarditis Other vasculitic disease Trauma	
	White spots	Trauma to the nail matrix	
	White transverse bands	Heavy metal poisoning	
	White/brown half and half nails	Chronic renal failure	
	White (leuconychia)	Hypoalbuminaemia (e.g. associated with cirrhosis)	
	Yellow	Psoriasis Fungal infection Jaundice Raynaud's disease Tetracycline Yellow nail syndrome (defective lymph drainage, nails grow very slowly, may be associated with pleural effusion)	Immunodeficiency Tuberculosis Bronchiectasis Sinusitis Nephritic syndrome
Brittle	Nails break easily—usually at the distal end	Effect of water and detergent, iron deficiency, hypothyroidism, digital ischaemia	

(*Continued*)

Table 16.4 *(Contd.)*

Change	Description of nail	Differential diagnosis
Clubbing ⚠ Refer any patient with unexplained clubbing for urgent CXR[N]	Loss of angle between nail fold and plate Bulbous finger tip Nail fold feels boggy	Respiratory: bronchial carcinoma (not small cell); chronic infection; fibrosing alveolitis; asbestosis Cardiac: SBE; congenital cyanotic heart disease Other: inflammatory bowel disease (Crohn's > UC); thyrotoxicosis; biliary cirrhosis; congenital; AV malformation
Koilonychia	Spoon-shaped nails	Physiological in children (especially big toenails) Congenital (most common) Other causes: iron deficiency anaemia; lichen planus; repeated exposure to detergents; haemochromatosis; Raynaud's disease; DM
Onycholysis	Separation of the nail from the nail bed	Psoriasis Fungal infection Trauma Thyrotoxicosis Tetracyclines Light (photo-onycholysis e.g. 2° to doxycycline use)
Pitting	Fine or coarse pits in the nail bed	Psoriasis Eczema Alopecia areata Lichen planus
Beau's lines	Transverse grooves	Any severe illness which affects growth of the nail matrix
Ridging	Transverse	Beau's lines Eczema Psoriasis Chronic paronychia Habit tic dystrophy (thumb > other fingernails—due to habitual rubbing/picking at the cuticle)
	Longitudinal	Lichen planus Raynaud's disease Darier's disease (keratosis follicularis—genetic disorder appearing in adolescence mainly affecting skin/nails)
Nail fold telangiectasia	Dilated capillaries and nail fold erythema	Normal change of ageing Connective tissue disorders
Tumours of the nail fold	Benign	Viral warts Myxoid (mucus) cysts (treat with steroid injection, cryotherapy or surgical excision) Periungual fibroma (associated with tuberous sclerosis—appear at puberty)
	Malignant	Melanoma Squamous cell carcinoma

Further information

Primary Care Dermatology Society Nail disorders. 🔗 www.pcds.org.uk/clinical-guidance/nails

Atopic eczema

Affects 15–20% of schoolchildren and 2–10% of adults—usually starts <6mo of age, and by 1y 60% of those likely to develop eczema will have done so. Associated with the filaggrin gene mutation and other atopic conditions. Remission occurs by 15y of age in 75%, although some relapse later.

Differential diagnosis Scabies; ringworm; rare syndromes e.g. Wiskott–Aldrich syndrome; dermatitis herpetiformis.

Presentation Waxing/waning itchy condition. ❗ If no itching, eczema is unlikely.

Infants Presentation varies in severity—from minor roughened patches of skin to itchy, vesicular, exudative eczema on face (Figure 16.8), trunk ± hands ± 2° infection. May cause sleep disturbance. >½ are free of eczema by 18mo.

Children >18mo Involves antecubital and popliteal fossae (Figure 16.9), neck, wrists, and ankles. Lichenification, excoriation, and dry skin are common. Face may be erythematous and have typical infraorbital folds. Loss of self-esteem and behaviour and sleep problems are common.

Adults Most commonly irritant hand dermatitis (Figure 16.10) in a person with past history of atopic eczema. A few continue to have generalized atopic eczema; <2% develop new eczema aged >20y. May interfere with employment/social activities. Exacerbated by stress.

Diagnosis Itchy skin *plus* ≥3 of:

- Itching in skin creases
- History of asthma or hay fever
- Onset in the first 2y of life
- Generally dry skin
- Visible flexural eczema

Assessment Ask about:

- Family (two-thirds) and personal history of atopy and eczema
- Onset and distribution of the disease
- Aggravating factors (pets, irritants e.g. soaps/detergents, allergens)
- Sleep disturbance due to itching/rubbing
- Impact on quality of life (school work, career, social life)
- Previous treatments (including dietary restrictions), expectations of treatment, and other medications being taken (e.g. steroids for asthma)

⚠ **Nipple eczema** May be Paget's disease of the breast (➔ p. 674). Refer urgently to breast surgeon if no response to topical treatment[N].

Complications

- Skin thickening and scaling
- Bacterial infection 2° infection (usually with *Staph. aureus*) commonly causes exacerbations and may not be obvious. Suggested by presence of crusting or weeping, or sudden deterioration of eczema
- Viral infection ↑ susceptibility to infection, e.g. viral warts, molluscum. *Eczema herpeticum*—propensity to develop widespread lesions with HSV and varicella zoster virus—may require admission and IV aciclovir
- Cataracts Rarely occur in young adults with very severe eczema
- Growth retardation Children with severe eczema, cause unknown. Keep a growth chart for children with chronic severe eczema

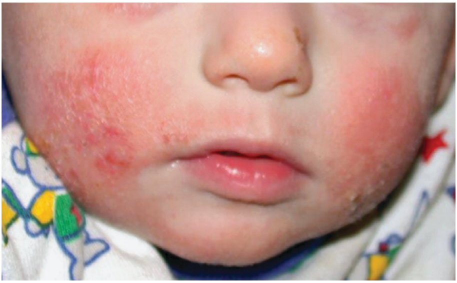

Figure 16.8 Infantile eczema on the cheeks

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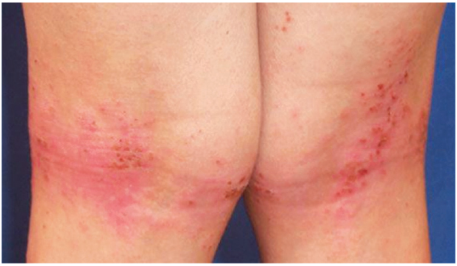

Figure 16.9 Childhood eczema involving both popliteal fossae

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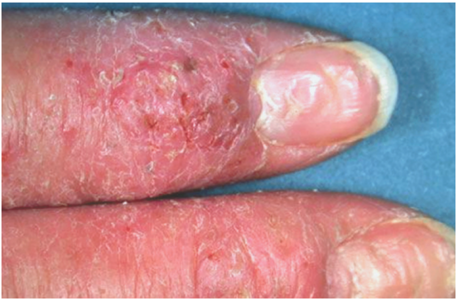

Figure 16.10 Hand (contact) dermatitis in an adult

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Management

- Education—explain the condition and provide verbal and written information on a stepped approach to care and management of flares^{N}
- Advise—loose cotton clothing; avoid wool (exacerbates eczema); avoid excessive heat; keep nails short; gloves in bed
- If a specific irritant is identified (e.g. house dust mite, pets) then avoid

Specific treatment

- Emollients Topical creams/ointments and bath emollients—use regularly on skin and as soap substitutes—even if skin is clear. May need to try several to find one that suits. Ideally apply 3–4×/d to moist skin. Ensure enough is supplied. Addition of an antipruritic, e.g. lauromacrogol, to the emollient may help break the scratch–itch cycle. Addition of an antiseptic to bath emollient may ↓ bacterial infection
- Topical steroids Prescribe the least potent strength that is effective. Use od or bd. Ointments are preferable on dry eczema; creams on wet, exudative eczema. Recurrent flares—once flare settles consider 2× weekly topical steroids as maintenance
- Antibiotics e.g. flucloxacillin 250–500mg qds for 7d—only use if a flare is not responding to ↑ topical steroid regimen. Swab if antibiotic treatment is ineffective
- Oral steroids Rescue therapy while waiting for an urgent consultant opinion. Only use short courses, e.g. prednisolone 20–30mg od for 5d
- Topical immunosuppressants e.g. pimecrolimus cream or tacrolimus ointment
- Antihistamines Short-course sedative antihistamines given nocte ↓ desire to itch, e.g. promethazine, hydroxyzine
- Bandages Excoriated or lichenified eczema—ichthammol (e.g. Ichthaband®) or zinc and calamine (e.g. Calaband®). Bandages can be applied at night on top of steroid ointment. Refer to dermatology
- Wet wrapping Used for exudative eczema—elasticated tubular bandage or tubular gauze soaked in emollient is applied and covered with a dry bandage. Refer to dermatology
- Dietary manipulation Few (<10%) benefit. Egg and milk are most commonly excluded. Refer to a dietician to avoid malnutrition

Referral *E* = Emergency admission; *U* = Urgent; *S* = Soon; *R* = Routine.

- Infection with disseminated HSV (eczema herpeticum)—*E*
- Severe eczema resistant to treatment. Additional secondary care treatments include phototherapy and immunosuppressive agents—*U*
- Infection which cannot be cleared in primary care—*U*
- Severe social/psychological problems due to eczema—*S*
- Treatment requires excessive amounts of topical steroids—*S*
- Failure to control symptoms in primary care—*R*
- Patient/family might benefit from additional advice on application of treatments (e.g. bandaging techniques)—*R*
- Patch testing required if contact dermatitis suspected—*R*
- Dietary factors are suspected (refer direct to dietician)—*R*

Prurigo nodularis (nodular prurigo) Intensely itchy firm lumps 1–2cm in diameter. Cause unknown, but 80% have a history of atopy. Exclude scabies and systemic causes of itching (➔ p. 566). *Treatment*: topical steroids ± occlusion. May need sedating antihistamine for itch.

Contact dermatitis Precipitated by an exogenous agent which is:

- **Irritant** (e.g. water, abrasives, chemicals, detergent) *or*
- **Allergen** (e.g. nickel—10% 5, 1% ♂; chrome; rubber)

Clinical presentation is often indistinguishable. More common in patients with a past history of atopic eczema. In some patients contact dermatitis may be an industrial disease (➲ p. 90). *Differential diagnosis*: endogenous eczema, psoriasis, fungal infection.

Presentation Affects any part of the body—most commonly the hands. Site and knowledge of occupation, hobbies, sports, etc. suggest cause.

- **Acute** Itchy erythema and skin oedema ± papules, vesicles, or blisters
- **Chronic** Lichenification, scaling, and fissuring

Management

- **Identification of the allergen or irritant** Consider referral for patch testing (➲ p. 660)
- **Exclusion of allergen or irritant from the environment**—if possible. There is some evidence that nickel avoidance diets can help patients with nickel sensitivity[G]. Nickel testing kits are available from dermatology departments
- **Hand care** Table 16.5
- **Emollients** Help skin to recover—apply frequently
- **Topical steroids** Help but are secondary to avoidance measures
- **Exclude/treat secondary infection**

Table 16.5 Hand care

Hand washing	Use warm water and substitute soap with emollient, e.g. aqueous cream; dry with a clean cotton towel—avoid paper towels or drying machines
Avoidance	Avoid handling hair preparations (including shampoos), other detergents, household or industrial cleaning fluids, raw vegetables (e.g. peeling potatoes, tomato juice); fruits (e.g. peeling oranges); or raw meat
Protection	If performing any task where hands would get wet, or any of the substances listed previously in table are being handled, wear cotton gloves under PVC gloves. Wear gloves for dusty work or in the cold
Medication	Use emollients frequently throughout the day (e.g. Diprobase® cream). If necessary, apply a thin layer of steroid ointment od/bd

Further information

NICE (2007) Atopic eczema in children under 12. 🖱 www.nice.org.uk/guidance/cg57

Primary Care Dermatology Society

- Atopic eczema (2019). 🖱 www.pcds.org.uk/clinical-guidance/atopic-eczema
- Contact allergic dermatitis (2019). 🖱 www.pcds.org.uk/clinical-guidance/eczema-contact-allergic-dermatitis-including-latex-and-rubber-allergy
- Hand (and foot) eczema (2019). 🖱 www.pcds.org.uk/clinical-guidance/eczema-hand-dermatitis

Patient information and support

National Eczema Society ☎ 0800 089 1122 🖱 www.eczema.org

Other eczemas

Discoid (nummular) eczema

- Middle-aged/elderly patients. ♂ > ♀. Unknown cause
- Presentation Intensely itchy, coin-shaped lesions on limbs. Tend to be symmetrical. May be vesicular or chronic and lichenified
- Differential diagnosis Tinea corporis; contact dermatitis, psoriasis
- Management Often clears spontaneously after a few weeks but tends to recur. If treatment is needed, use a moderate or potent topical steroid. Secondary infection is common—treat with topical/systemic antibiotics. A sedating antihistamine (e.g. hydroxyzine 50mg nocte) may be useful if sleep is disturbed by itching

Venous (stasis, varicose) eczema

- Middle-aged/elderly patients. ♀ > ♂
- Associated with underlying venous disease
- Early signs Capillary veins and haemosiderin deposition around the ankles and over prominent varicose veins
- Later signs Figure 16.11. Eczema ± lipodermatosclerosis (fibrosis of the dermis and subcutaneous tissue) ± ulceration
- Management Emollients ± mild or moderate steroid ointment (avoid long-term use) and compression hosiery. Treat venous disease (➔ p. 258) or ulceration (➔ p. 584) on its own merits

Pompholyx Sago-like intensely itchy vesicles on the sides of fingers ± palms/soles. No associated atopic eczema or contact dermatitis. Young adults. More common in warm weather. Frequently recurrent. Treat with emollients and topical steroids (some need potent steroids). Treat any infection with oral antibiotics. In severe cases refer to dermatology for wet dressings.

Lichen simplex chronicus Area of lichenified eczema due to repeated rubbing/scratching. May be due to habit or stress. *Treatment*: take scrapings to exclude fungal infection, topical steroids, occlusive dressing (e.g. Duoderm®) to protect skin from scratching.

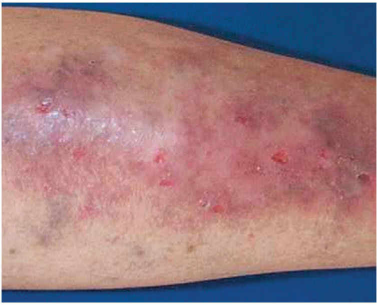

Figure 16.11 Varicose eczema showing haemosiderin deposition, excoriated eczematous lesions, and lipodermatosclerosis

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Asteatotic eczema (eczema craquelé)

- **Risk factors** ↑ age; overwashing; dry climate; hypothyroidism; diuretics
- **Presentation** Dry, itchy eczema with fine, crazy-paving pattern of fissuring and cracking of the skin of the limbs
- **Management** Treat with emollients ± mild topical steroid

Seborrhoeic dermatitis Chronic scaly eruption affecting scalp, face, and/or chest. *Differential diagnosis*: psoriasis, rosacea, contact dermatitis, fungal infection. 5 patterns:

- **Scalp and facial involvement** Most common in young men. Excessive dandruff, itchy scaly erythematous eruption affecting sides of the nose, eyes, ears, hairline. May be associated blepharitis
- **Petaloid** Dry, scaly eczema over the pre-sternal area
- ***Malassezia* (*Pityrosporum*) folliculitis** Erythematous follicular eruption with papules/pustules over the back
- **Flexural** Most common in the elderly. Axillae, groins, and submammary areas. Moist intertrigo. Associated with 2° candidal infection
- **Infantile seborrhoeic eczema** ➔ p. 883

Treatment

- **Facial, truncal, and flexural involvement** Ketoconazole 2% shampoo (2×/wk for 2–4wk, rinse off after 3–5min) or clotrimazole/miconazole cream ± hydrocortisone. Consider itraconazole 200mg od for 7d or fluconazole 50mg od for 2wk if not clearing
- **Scalp lesions** Ketoconazole 2% or coal tar shampoo. If resistant, apply 2% sulphur + 2% salicylic acid cream several hours before shampooing
- **Recurrence** requiring repeated treatment is common. Consider maintenance treatment with topical antifungal every other week

❗ Severe or recalcitrant seborrhoeic dermatitis is an indicator disease for HIV infection—offer HIV testing[N].

Dandruff Exaggerated physiological exfoliation of fine scales from an otherwise normal scalp. More severe forms merge with seborrhoeic dermatitis and treatment is the same.

Further information

British HIV Association, BASHH, and British Infection Society UK national guidelines for HIV testing. 🖱 www.bhiva.org/HIV-testing-guidelines.aspx

Primary Care Dermatology Society

- Asteatotic eczema (2014). 🖱 www.pcds.org.uk/clinical-guidance/eczema-asteatotic-eczema-syn.-xerotic-eczema-eczema-craquele
- Discoid eczema (2016). 🖱 www.pcds.org.uk/clinical-guidance/eczema-discoid-syn.-nummular-eczema
- Lichen simplex (2019). 🖱 www.pcds.org.uk/clinical-guidance/lichen-simplex-chronicus
- Seborrhoeic dermatitis (2016). 🖱 www.pcds.org.uk/clinical-guidance/seborrhoeic-eczema
- Varicose eczema (2016). 🖱 www.pcds.org.uk/clinical-guidance/eczema-gravitational-eczema-syn.-varicose-eczema-or-stasis-dermatitis

Patient information and support

National Eczema Society ☎ 0800 089 1122 🖱 www.eczema.org

Ulcers

Leg ulcer Painful and debilitating condition affecting 1% of the adult population and ~4% of those >65y. *Common causes*:

- Venous (70–80%)—above the medial/lateral malleolus of the ankle
- Mixed venous/arterial (10–20%)
- Arterial (10%)—on the shin, toes, or over pressure points (under the heel or over malleoli)
- Neuropathic—on the sole of the foot or over pressure points

Rarer causes Trauma; obesity; immobility; vasculitis (RA, SLE, PAN); malignancy; osteomyelitis; blood dyscrasias; lymphoedema; self-harm.

History Duration of ulceration; pain (painful unless neuropathic when often painless); mobility; drugs (causing ulcers, e.g. nicorandil, or impeding healing, e.g. β-blockers, immunosuppressants, NSAIDs); past history of ulceration, DVT, or varicose vein surgery; history of trauma to the limb; systemic disease (e.g. DM, peripheral vascular disease, RA).

Examination *Ulcer*: position; size; depth; evidence of infection; surrounding callus (typical of neuropathic ulcers). *Leg*: pulses; varicose veins and/or signs of venous hypertension—haemosiderin pigmentation, varicose eczema, atrophie blanche (white lacy scars), lipodermatosclerosis; sensation (↓ when peripheral neuropathy); range of joint movement.

Investigation

- Bloods—FBC, ESR, FBG/HbA1c, LFTs, U&E/eGFR, TFTs, lipid profile
- Ankle–brachial pressure index (ABPI; ➲ p. 209)
- Swab for M,C&S if any signs of cellulitis/infection
- Diabetic ulcer—if infection, X-ray foot to exclude osteomyelitis

Management ❗ Refer diabetic foot ulceration to foot clinic—➲ p. 330.

- Apply dressings—choice depends on where the ulcer is in the healing process, whether it is infected, and amount of exudate
- Depending on ABPI, consider compression therapy and/or referral for vascular assessment (Table 16.6). With multilayer compression, ~70% of venous ulcers heal in <3mo. If not improving in 3mo, or not healed by 12mo, refer to the tissue viability service or dermatology

Complications

- Infection—treat with systemic antibiotics if advancing ulcer edge, cellulitis, or systemic symptoms. Antibiotic choice is guided by swab results
- Overgranulation—treat with silver nitrate cautery or potent topical steroid, e.g. Dermovate®
- Contact dermatitis to topical medicaments and dressings. Treat with topical steroid and consider referral for patch testing if suspected
- Malignant change—squamous cell cancer (rare). Refer urgently (to be seen in <2wk) for biopsy to confirm diagnosis

Prevention of recurrence 5y recurrence rate is 40%—graduated compression hosiery ↓ recurrence. Below-knee class 2 stockings are adequate and can be prescribed with NHS prescription. They can be difficult to apply, especially with arthritic hands. Applicators are available via NHS prescription. Consider referral for vascular surgery review to assess whether vein surgery may ↓ recurrence.

Table 16.6 ABPI and leg ulcer management

ABPI	Description	Management
>1.2	Calcification of vessels is likely	Refer for vascular assessment
0.8–1.2	Suggests venous disease	Suitable for full compression therapy unless DM or RA[a]
0.6–0.8	Suggests mixed vascular disease	Consider referral for vascular assessment; most can be managed with reduced compression therapy
<0.6	Suggests arterial disease	Refer for vascular assessment. Not suitable for compression therapy

[a] ABPI can be falsely high in RA and DM—extra care is needed when using compression therapy. Consider early referral for specialist advice.

Pyoderma gangrenosum Uncommon cause of ulceration. Starts as a pustule/inflamed nodule that breaks down to form an ulcer that often expands rapidly and is painful (Figure 16.12). The ulcer has a purplish margin and surrounding erythema. Usually on trunk/lower limbs. Refer to dermatology. *Causes*: UC (50% patients with pyoderma gangrenosum); Crohn's disease; RA; Behçet's syndrome; multiple myeloma and monoclonal gammopathy; leukaemia.

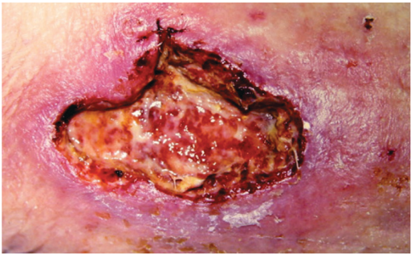

Figure 16.12 Pyoderma gangrenosum

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Bed sores or pressure ulcers

- Caused by pressure necrosis of the skin. Immobile patients are at high risk—especially if frail ± incontinent
- If at risk, refer to the DN for advice on prevention—protective mattress/cushions, incontinence, guidance on positioning/movement
- Warn carers to make contact with the DN if a red patch does not improve 24h after relieving the pressure on the area. Treat aggressively and admit if not resolving

Further information

NICE (2014) Pressure ulcers. 🖱 www.nice.org.uk/guidance/cg179

Primary Care Dermatology Society (2016) Leg ulcers and disorders of venous insufficiency. 🖱 www.pcds.org.uk/clinical-guidance/leg-ulcers

SIGN (2010) Management of chronic venous leg ulcers. 🖱 www.sign.ac.uk/assets/sign120.pdf

Urticaria and angio-oedema

Anaphylaxis ➔ p. 1054

Urticaria (hives or nettle rash) Common; affects 1 in 6 at some time. Superficial, itchy swellings of the skin or *weals* (Figure 16.13) come and go in an attack giving the appearance of a shifting rash.

Angio-oedema Deeper longer-lasting swellings; painful rather than itchy. Commonly affect eyes, lips, genitalia, hands, and/or feet. May affect bowel (abdominal pain, nausea, vomiting, diarrhoea) or airway (tongue swelling, shortness of breath, wheeze—consider anaphylaxis ➔ p. 1054).

Classification Table 16.7. Half present with urticaria alone; 1 in 10 has angio-oedema alone; the remainder have both.

Management of acute urticaria Only treat if needed:

- Try antihistamines for itch—non-sedating for daytime (e.g. cetirizine, fexofenadine) ± sedative if interferes with sleep (e.g. chlorphenamine, hydroxyzine). Many dermatologists use up to 2× standard dose (off-label use). If one antihistamine is ineffective, try another
- Topical menthol 1% cream is an alternative/adjunct to antihistamines
- If severe, consider short-course steroids (e.g. prednisolone 40mg od for 3–5d). If rebound symptoms after stopping, seek specialist advice

Management of chronic urticaria Check FBC, ESR, and TFTs. Assess severity and impact of symptoms. Identify potential causes. Advise to avoid non-specific aggravating factors, e.g. overheating, stress, alcohol, aspirin/codeine, and NSAIDs if aspirin sensitive. Prescribe antihistamines as for acute urticaria. If these measures do not control symptoms, refer. Other treatments are usually specialist initiated and unlicensed. They include H_2 receptor antagonists (e.g. cimetidine, ranitidine) and anti-leukotrienes (e.g. montelukast)—response is highly variable.

Management of angio-oedema

- If anaphylaxis is suspected, give adrenaline and admit (➔ p. 1054)
- If any airway compromise, admit—even if anaphylaxis is not suspected
- Otherwise treat as for acute urticaria; monitor for airway compromise
- If not taking ACE inhibitor, refer to allergy clinic/immunology; if taking ACE inhibitor, stop—refer if symptoms continue/recur after >3mo

Urticaria pigmentosa (cutaneous mastocytosis) Appears in infancy (usually <2wk old). Dark freckle-like lesions on the face, limbs, or trunk become urticarial when the skin is rubbed. No treatment is needed—clears spontaneously in childhood.

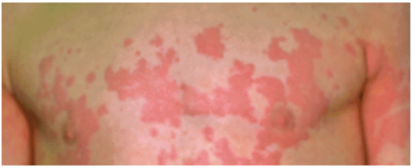

Figure 16.13 Typical urticarial lesions in a child

Table 16.7 Classification of urticaria and angio-oedema

Type	Features
Ordinary/ idiopathic	Spontaneous weals ± angio-oedema. Individual weals last 2–24h. • Acute <6wk of continuous activity • Chronic ≥6wk of continuous activity—affects 1–5/1000 May remit/relapse. Relapses are triggered by illness, stress, drugs, alcohol, or hormonal changes (e.g. menstruation). 50% resolve in 3–5y; 20% persist >10y. Associated with autoimmune thyroid disease (↑ 2×); children/adolescents have ↑ prevalence of coeliac disease. Severe impact on quality of life—14% develop depression • Episodic (acute intermittent/recurrent) Symptoms last hours/days but recur over months/years. Treated like chronic urticaria *Triggers include*: stress, overheating, drugs, alcohol, viral infections
Physical	Except for delayed pressure urticaria, weals last <1h. Induced by a specific physical stimulus; avoidance prevents attacks • Mechanical Delayed pressure urticaria (weals appear in 2–6h and fade over 48h); symptomatic dermographism; vibratory angio-oedema • Thermal Cholinergic urticaria (induced by sweating); cold contact urticaria; localized heat urticaria • Other Aquagenic urticaria (contact with water); solar urticaria; exercise-induced anaphylaxis
Contact	Weals last <2h. Caused by allergens (e.g. nuts, shellfish, milk, eggs, penicillin, insect stings, latex) or chemicals (e.g. drugs—opioids, aspirin, NSAIDs; radio-contrast media; food additives—azo-dyes, preservatives). Refer for allergy testing if specific allergen is suspected
Urticarial vasculitis	Individual weals last >24h; lesions are burning/painful rather than itchy; and/or lesions leave scaling, bruising, purpura/petechial haemorrhages. Suspect if relentless rather than self-limiting urticaria. *Other features*: joint pains, fever and/or malaise. Refer. Diagnosis is confirmed by skin biopsy. *Specialist treatment*: steroids and/or immunosuppressive agents
Autoimmune	Urticaria, pyrexia, and malaise + disease-specific features. May be hereditary or acquired (e.g. SLE)—treat the cause
Lone angio-oedema	Swellings last <3d. *Causes*: idiopathic; drug-induced (ACE inhibitors, ARBs, NSAIDs); C1 esterase inhibitor deficiency

C_1 esterase inhibitor deficiency (hereditary angio-oedema)

Autosomal dominant—usually presents in puberty with episodes of angio-oedema without weals. ↓ C4 level suggests the diagnosis. Emergency treatment is with hospital admission for C1 inhibitor concentrate infusion. Maintenance therapy (under consultant supervision with anabolic steroids or tranexamic acid) is necessary only for patients with symptomatic recurring angio-oedema or related abdominal pain.

Further information

British Association of Dermatologists (2007) Guidelines for evaluation and management of urticaria in adults and children. 🔗 https://onlinelibrary.wiley.com/doi/full/10.1111/j.1365-2133.2007.08283.x

BSACI (2015) Guidelines for the management of chronic urticaria and angioedema. 🔗 http://onlinelibrary.wiley.com/doi/10.1111/cea.12494/epdf

Primary Care Dermatology Society (2017) Urticaria and angioedema—overview. 🔗 www.pcds.org.uk/clinical-guidance/urticaria-and-angioedema

Acne

Chronic inflammatory condition characterized by comedones, papules, pustules, cysts, and scars. Acne vulgaris is common and affects >80% teenagers; 50% have a family history. *Peak age*: 18y; ♂ = ♀.

Cause Complex—androgen secretion results in ↑ sebum excretion; pilosebaceous duct blockage (producing comedones); colonization of the duct with *Propionibacterium acnes* bacteria and release of inflammatory mediators. Inflammatory acne is the result of the host response to the follicular *Propionibacterium acnes*.

Rarer causes

- Endocrine—PCOS, Cushing's, virilizing tumours
- Squeezing—*acne excoriée*
- Aromatic industrial chemicals—*chloracne*
- Cosmetics
- Drugs—systemic steroids, androgens, topical steroids
- Infantile—faces of male infants—cause unknown
- Physical occlusion, e.g. under a violinist's chin

Presentation Spots on face, neck, ± back and chest. Examination reveals blackheads (dilated pores with black plug of keratin = comedones) and whiteheads (small cream-coloured dome-shaped papules); red papules; pustules ± cysts. There may be scarring from old lesions. Burrowing abscesses and sinuses with scarring (*conglobate acne*) are seen in severe cases. Scars may become keloidal. ❗ Severity of acne is often overestimated by the patient and minimized by the doctor.

Differential diagnosis Rosacea (➔ p. 570), bacterial folliculitis (often coexists—➔ p. 607), milia (➔ p. 595), perioral dermatitis.

Management *Aims to*: ↓ number of lesions, prevent scarring, and ↓ the psychological impact of the condition.

- **Misconceptions** Explain:
  - Acne is not a disease of poor hygiene and is not infectious. The black tip of a comedone is oxidized sebum not dirt
  - 💣 A high glycaemic index diet may be associated with acne: suggest ↓ sugar and ↑ wholegrains and fresh fruit/vegetables
  - Picking at acne does not improve it and can cause scarring
- **General measures** Encourage to wash no more than 2×/d using a fragrance-free cleanser. Recommend non-comedogenic make-up (avoid oil-based cosmetics) and emollients

Medication Table 16.8. Warn patients any treatment takes up to 8wk to work and may irritate the skin initially. Topical and systemic antibiotics should not be prescribed together, or used as sole treatment as antibiotic resistance is a growing concern. All treatments should be routinely reviewed after 12wk. ⚠ In the event of pregnancy all retinoids and oral tetracyclines should be discontinued.

Review If treatment goals are met after 12wk, discontinue topical/oral antibiotics; consider maintenance treatment with topical therapy. If treatment goals are *not* reached after 12wk, review treatment adherence; consider alternative treatment and/or referral.

Table 16.8 Acne treatment ladder

Step	Description	Management
Step 1	Comedonal acne	**First-line** Topical retinoid (e.g. isotretinoin, adapalene ± benzoyl peroxide). Start with low-strength preparation 2–3 nights/wk—gradually ↑ frequency/duration/strength of application—warn patients that treatment may dry and irritate the skin initially and to avoid the sun **Second-line** Topical azelaic acid (e.g. Skinoren®, Finacea®)
Step 2	Mild to moderate papular/pustular acne	Try a fixed-dose combination topical treatment: ● **First-line** Adapalene + benzoyl peroxide (Epiduo®) ● **Second-line** Clindamycin + benzoyl peroxide (Duac®) ● **Other options** Clindamycin + tretinoin (Treclin®); erythromycin combinations ❗ Warn patients that treatment may dry and irritate the skin initially and to avoid the sun
Step 3	Treatment failure or more extensive acne	Combine oral antibiotics with a topical agent (usually benzoyl peroxide but consider topical retinoid if unable to tolerate benzoyl peroxide) ● **First-line** Lymecycline 408mg od. Review after 6wk. If inadequate response, switch to doxycycline 100mg od (may cause photosensitive reaction) ● **Alternatives if pregnant or <12y** Erythromycin; trimethoprim Continue treatment for a maximum of 12wk. Continue topical treatment once antibiotics stopped. Course of antibiotics can be repeated if acne recurs
Step 4	Moderate to severe acne in ♀	Consider hormonal contraception with a 2nd/3rd-generation combined oral contraceptive pill. Co-cyprindiol is reserved for ♀ for whom other treatments have failed; discontinue 3mo after acne control is achieved

Complications Acne is not a trivial disease—it can cause scars (both skin and emotional) that last a lifetime. Anxiety, social isolation, and lack of self-confidence are common—ask about these factors; treat as needed.

Perioral dermatitis Papules and pustules which appear around the mouth and chin of a woman, often after use of topical steroids. Treat with oral tetracycline as for acne.

Referral to dermatology *U* = Urgent; *R* = Routine.

- Acne fulminans—seen in adolescent boys; severe acne is associated with fever, arthritis, and vasculitis—*U*
- Nodulo-cystic acne for consideration of oral retinoid (start oral antibiotics and refer immediately)—*U*
- Severe social/psychological sequelae—*U*
- At risk of/developing scarring despite primary care remedies—*R*
- Poor treatment response after >6mo—*R*
- Suspected underlying cause for acne (e.g. PCOS—➔ p. 700)—*R*
- Diagnostic uncertainty—*R*

Further information

Primary Care Dermatology Society (2019) Acne vulgaris. 🖱 www.pcds.org.uk/clinical-guidance/acne-vulgaris

Psoriasis

Chronic, non-infectious inflammatory skin condition. Epidermal cell proliferation rate is ↑ ×20 and turnover time ↓ from 28 to 4d. Affects ~3% Caucasian population (less in other races). Mean age 28y; rare <8y. ♂ = ♀.

Associations Inflammatory bowel disease (Crohn's > UC); ↑ risk CVD (check BP, lipids, exclude DM) and venous thromboembolism (➔ p. 260).

Cause Autoimmune disease. Genetic predisposition—FH is present in 40–50% (~75% if onset is at age <20y). If 1 parent is affected, there is a 28% probability that a child will be affected; 65% chance if 2 parents affected. Environmental factors trigger disease:

- Trauma (Koebner phenomenon)
- Infection
- Drugs (e.g. β-blockers, NSAIDs, lithium)
- Alcohol
- Pregnancy
- Psychological stress
- Sunlight—usually beneficial but aggravates psoriasis in ~10%

Presentation Two peaks of incidence—young adults and those aged 60–70y. Generally presents with skin changes but no other symptoms. Skin changes may be accompanied by itch.

Patterns of psoriasis Table 16.9

Assessment All patients with psoriasis should be assessed at least 1×/y and if any new/worsening symptoms. Consider:

- % of body surface affected
- Involvement of nails (~50%)
- Global disease severity rating from clinician's and patient's perspective: clear, nearly clear, mild, moderate, severe, or very severe
- Presence of arthritis—affects ~14–30%; if no arthritis, screen with the PEST questionnaire annually (Box 16.2)
- High-impact or difficult-to-treat sites (e.g. face, scalp, palms/soles, flexures, genitals)
- Impact of disease on physical, psychological, and social well-being; impact on patient, family, and carers; consider depression (➔ p. 173)
- Presence of co-morbidities (including CVD risk assessment—➔ p. 214)
- Systemic upset (e.g. fever, malaise)—may indicate unstable psoriasis requiring admission

Management of psoriasis ➔ p. 592

Box 16.2 Psoriasis Epidemiology Screening Tool (PEST)

- Have you ever had a swollen joint (or joints)?
- Has a doctor ever told you that you have arthritis?
- Do your fingernails or toenails have holes or pits?
- Have you had pain in your heel?
- Have you had a finger or toe that was completely swollen or painful for no apparent reason?

Score 1 point for each +ve answer. A score of ≥3 indicates high risk of psoriatic arthropathy and should prompt rheumatology referral.

❗ Does not detect axial arthritis or inflammatory back pain.

Source: data from Helliwell PS, Psoriasis Epidemiology Screening Tool (PEST). *J Rheumatol*. 2011; **38**(3):551–2.

Table 16.9 Patterns of psoriasis

Pattern	Features
Erythroderma	➔ p. 568—admit as a medical emergency
Generalized pustular	Rare but serious. Unwell with fever and malaise. Sheets of small, sterile, yellowish pustules develop on an erythematous background and spread rapidly. Admit as an emergency
Plaque (Figure 16.14a)	Most common form (90%). Well-defined disc-shaped plaques involving the knees, elbows, scalp, hair margin, or sacrum. Plaques are usually red and covered with waxy white scales which may leave bleeding points if detached. Plaques may be itchy *Differential diagnosis*: drug eruption; hypertrophic lichen planus
Scalp	Very common. May be confused with dandruff but generally better demarcated and thicker scales
Guttate (Figure 16.14b)	Acute symmetrical raindrop lesions on trunk/limbs. Most common in adolescents/young adults—may follow streptococcal throat infection. Usually clears in 2–3mo. *Differential diagnosis*: pityriasis rosea
Flexural	Affects axillae, submammary areas, and natal cleft. Plaques are smooth and often glazed. Most common in elderly patients. *Differential diagnosis*: flexural candidiasis
Nail (Figure 16.14c)	Nail bed is affected in 50%. Fingernails > toenails. Thimble pitting, onycholysis, and oily patches (oily, brownish-yellow discolouration of the nail bed—often adjacent to onycholysis). Associated with arthropathy. Treatment is difficult. *Differential diagnosis*: fungal nail infection (❗ can coexist so send clippings for mycology)
Palmoplantar pustulosis	Yellow/brown-coloured sterile pustules on palms or soles
Napkin	Well-defined eruption in nappy area of infants
Psoriatic arthropathy	~40% patients with skin changes. ♂ = ♀ Distal arthritis DIP joint swelling of hands/feet ± flexion deformity Rheumatoid-like Polyarthropathy similar to rheumatoid arthritis (➔ p. 488) but less symmetrical and rheumatoid factor is −ve. Mutilans Associated with severe psoriasis. Erosions in small bones of hands/feet ± spine. Bones dissolve → progressive deformity Ankylosing spondylitis/sacroiliitis Usually HLA B27 +ve; ➔ p. 492 Refer to rheumatology for confirmation of diagnosis, advice on management, and disease-modifying drugs

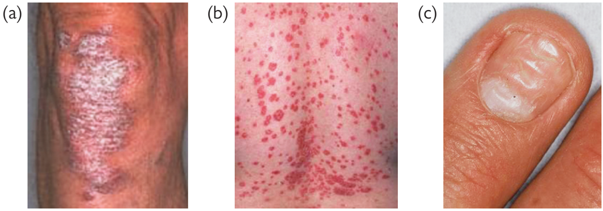

Figure 16.14 Psoriasis: (a) silvery scale of plaque psoriasis, (b) widespread rash of guttate psoriasis, and (c) nail changes in psoriasis

(a) and (b) Reproduced from www.psoriasisguide.ca with permission from Skin Information, SkinCareGuide.com

Management of psoriasis

Explain the condition/treatment options. Discuss the chronic nature of psoriasis and reassure not contagious. Provide information about psoriasis and advise on self-help organizations.

Assessment and regular review ➔ p. 590

Emollients Prescribe emollients to be used continuously, even when active treatment is not required. ↓ the amount of scale and itch.

Topical treatments Choice depends on extent and site of psoriasis:
- Cream, lotion, or gel is suitable for widespread psoriasis
- Lotion, solution, or gel is suitable for the scalp or hair-bearing areas
- Ointment is preferred to treat areas with thick adherent scale

Plaque psoriasis on the trunk/limbs Betamethasone/calcipotriol combination products (e.g. Dovobet® gel/ointment, Enstilar® foam) are used by many GPs as first-line treatment (💣 NICE recommends separate application). Discontinue when the skin feels smooth, even though still looking pink/red, or after 8wk. Do not restart a potent topical steroid for ≥4wk. Recommend ongoing treatment with an emollient ± vitamin D analogue (calcitriol or calcipotriol) as needed. *Alternatives include:*
- Tar preparation (e.g. Exorex® lotion) for large, thin plaques
- Topical retinoid (e.g. tazarotene)—co-prescribe a topical steroid (applied at a separate time) to ↓ skin irritation

❗ Dithranol should only be used under specialist supervision.

Scalp psoriasis Advise to use coal tar shampoo (e.g. Polytar®, Alphosyl®)—apply for ≥5min to allow time to penetrate scale before washing out. Benzalkonium chloride shampoo (Dermax®) is an alternative.

Treat flare-ups with topical steroid (e.g. betamethasone scalp application or clobetasol shampoo) or topical steroid/vitamin D analogue combination (e.g. Dovobet® gel). Do not wash the hair for 8–12h after application of gel or scalp application; leave shampoo on for ~20min before washing out. 1% hydrocortisone or clobetasone cream may be used to treat scalp margin psoriasis.

If thick scale, try a coal tar + salicylic acid preparation (e.g. Sebco®). Massage in for ~5min and then leave on for ≥2h (preferably overnight). Wash out with Capasal® shampoo. ❗ Warn patients treatment is messy and may need to cover scalp with a shower cap or old towel/pillow case.

Facial psoriasis Avoid potent topical steroids. Try vitamin D analogue (e.g. tacalcitol lotion/ointment, calcitriol ointment). Lower-strength topical steroids (e.g. 1% hydrocortisone or clobetasone cream) may be used but carry the risk of skin atrophy. An alternative is topical tacrolimus 0.1% ointment (off-label indication) if inadequate response.

Genital/flexural psoriasis Consider weaker steroids (e.g. 1% hydrocortisone or clobetasone cream) or vitamin D analogues (e.g. tacalcitol lotion/ointment, calcitriol ointment). Topical tacrolimus 0.1% ointment or pimecrolimus cream may be alternatives if inadequate response (both off-label indications). More potent steroid can be used in the gluteal cleft.

Hand/foot psoriasis

- If hyperkeratotic—try emollient + Diprosalic® ointment. If inadequate response, a stronger salicylic acid preparation may be required, e.g. 5% salicylic acid in yellow soft paraffin bd until the scale thins (❗ this is made up by pharmacies as a 'special' and may be very expensive)
- Pustular psoriasis—try potent topical steroid (e.g. clobetasol propionate 0.05% ointment, betamethasone 0.1% ointment). Application with occlusion (e.g. with cling film) may ↑ effect

Nail psoriasis Treatment is difficult. If distal nail involvement, cut the nail as short as possible and apply potent topical steroid or steroid/vitamin D analogue combination (e.g. Dovobet® ointment) od for 3mo. Nail varnish can be useful to disguise nail changes. Refer if nail changes are having major functional/cosmetic impact.

Guttate psoriasis Usually clears naturally in 2–3mo. Try emollients ± coal tar preparations (e.g. Exorex® lotion, Alphosyl-HC® cream) ± vitamin D analogue (applied directly to the lesions). If widespread/unresponsive, refer to dermatology for UVB phototherapy.

Review 4wk after starting a new topical treatment.

- If inadequate response and adherent to treatment, discuss the next treatment option
- If responding to treatment, advise to continue until skin is clear/nearly clear or up to the maximum treatment period (8wk for potent topical steroids). Relapse once treatment is stopped is likely; provide a supply of topical treatment to use as needed to maintain disease control

Referral *E* = Emergency; *U* = Urgent; *S* = Soon; *SR* = Routine.

- Unstable psoriasis—generalized pustular or eythrodermic psoriasis—*E*
- Acute guttate psoriasis requiring phototherapy—*U*
- All children <16y—*U/S/R*
- Immediately if topical therapy alone is unlikely to control symptoms (e.g. extensive—>10% body surface area; moderate/severe; nail disease with functional/cosmetic impact)—*S/R*
- Severe physical/social/psychological sequelae—*S*
- For management of associated arthropathy (to rheumatology)—*S*
- Diagnostic uncertainty—*S/R*
- Inadequate response to primary care treatments—*R*

Secondary care treatment options Phototherapy and PUVA; oral retinoids; cytotoxic, immunosuppressive, and biologic therapy; specialist nursing services.

Further information

NICE (2012, updated 2017) Psoriasis: assessment and management. ✆ www.nice.org.uk/guidance/cg153

Primary Care Dermatology Society (2019) Psoriasis: an overview. ✆ www.pcds.org.uk/clinical-guidance/psoriasis-an-overview

SIGN (2010) Diagnosis and management of psoriasis and psoriatic arthritis. ✆ www.sign.ac.uk/assets/sign121.pdf

Patient information and support

Psoriasis Association ✆ www.psoriasis-association.org.uk

Lichen planus and keratinization disorders

Lichen planus Very itchy, polygonal, flat-topped papular lesions 2–5mm diameter affecting flexor surfaces, palms/soles, mucous membranes (two-thirds), and genitalia in a symmetrical pattern. *Koebner phenomenon* (lesions occur in the line of damaged skin due to a scratch—Figure 16.15a) is exhibited. Papules may have a surface network of white lines (Wickham's striae). Initially papules are red but become violaceous. Papules flatten over a few months to leave pigmentation or occasionally become hypertrophic. Two-thirds of cases occur in the 30–60y age group. ♂ = ♀. Cause unknown. *Variants*:

- **Annular** (10%) Commonly on glans penis
- **Atrophic** Rare, associated with hypertrophic lesions
- **Bullous** Blistering is rare
- **Follicular** May occur with typical lichen planus or just affect the scalp
- **Hypertrophic** Plaques may persist for years
- **Mucous membrane** (Figure 16.15b) Alone or with skin changes

Differential diagnosis Lichenoid drug eruption; psoriasis.

Complications

- **Nail involvement** (10%) Longitudinal pitting and grooving
- **Scalp** Scarring alopecia
- **Malignant change** 💣 It is controversial whether oral lichen planus can undergo malignant transformation. If it can, risk is low (<2% over 10y). NICE recommends patients with confirmed oral lichen planus are monitored for oral cancer as part of routine dental examination[N]

Management Emollients and moderate/high-potency topical steroids provide symptomatic relief. Sedating antihistamines may be useful if sleep disturbed. Oral lesions can be treated with hydrocortisone pellets. Rarely, oral lesions are a reaction to mercury amalgam fillings—removing the fillings and replacing them with other materials may solve the problem. Refer to dermatology if:

- Diagnosis is in doubt
- Extensive involvement
- Potentially scarring nail dystrophy
- Resistant to topical treatment

Specialist treatment involves oral steroids ± PUVA.

Prognosis 50% are clear in <9mo; 15% have continuing symptoms >18mo; 20% have a further attack.

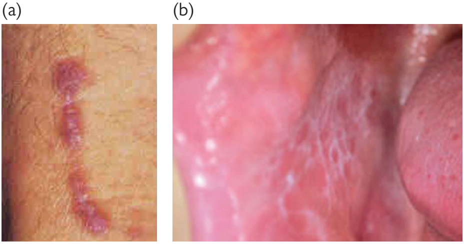

Figure 16.15 Lichen planus: (a) Koebner phenomenon and (b) oral lesions

Figure 16.15(b) reproduced with permission from New Zealand Dermatological Society Incorporated. Published online at: www.dermnetnz.org

Lichen planus-like drug eruptions Recorded after treatment with:

- Thiazide diuretics
- ACE inhibitors
- Tolbutamide
- Penicillamine
- Phenothiazines
- Streptomycin
- Tetracycline
- Isoniazid
- Gold
- Quinine
- Chloroquine

Resolution after withdrawal of drug is often slow.

Lichen sclerosus (hypoplastic vulval dystrophy) ➔ p. 707

Callosities Painless localized thickenings of the keratin layer—protective response to friction/pressure. *Management*: keratolytics, e.g. 5–10% salicylic acid ointment or 10% urea cream; attention to footwear.

Corns Painful. Develop at areas of high local pressure on the feet (e.g. where shoes press on bony protrusions). *Management*: attention to footwear, keratolytics, cushioning (e.g. corn pads). Occasionally surgery may be indicated if deformity of the foot causes recurrent corns.

Keratosis pilaris Common, sometimes inherited condition. Small horny plugs are on the upper thigh, upper arm, and face (Figure 16.16). Associated with ichthyosis vulgaris. Keratolytics, e.g. 10% urea cream, improve symptoms.

Ichthyosis Inherited disorders characterized by dry scaly skin. Most common form is *ichthyosis vulgaris*: prevalence 1 in 300, autosomal dominant. Small branny scales on extensor aspects of limbs and back. Mild and often undiagnosed. *Management*: topical emollients. Severe cases require dermatology advice.

Keratoderma Hyperkeratosis of palms and soles. *Tylosis* is diffuse hyperkeratosis of the palms and soles. It is usually inherited (autosomal dominant) but rarely may be associated with oesophageal cancer. Acquired keratoderma occurs in women around the menopause and patients with lichen planus. Treat with keratolytics, e.g. 10% urea cream.

Milia Small, white, raised 1–2mm diameter spots (Figure 16.17) resulting from small keratin-filled cysts. Usually on the face (upper cheeks and eyelids). Most common in children but can occur at any age. No treatment is required.

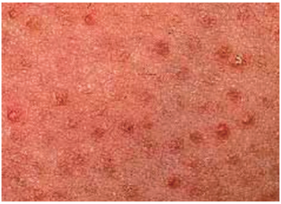

Figure 16.16 Keratosis pilaris

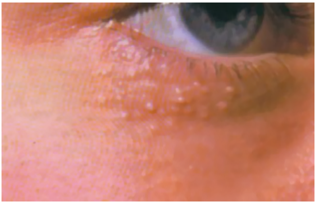

Figure 16.17 Milia

Further information

Primary Care Dermatology Society (2018) Lichen planus. 🖱 www.pcds.org.uk/clinical-guidance/lichen-planus

Pityriasis and seborrhoeic keratoses

Pityriasis rosea Acute self-limiting disorder most commonly affecting teenagers and young adults. *Cause*: unknown, possibly viral. Generalized eruption is preceded by the herald patch—a single large, oval lesion 2–5cm diameter (Figure 16.18). Several days later the rash appears consisting of many smaller lesions mainly on trunk (following the ribs giving a 'Christmas tree' pattern) but also upper arms and thighs. Lesions are oval, pink, and have a delicate 'collarette' of scale. May be asymptomatic or cause mild/moderate itch. Treatment does not speed clearance. Topical steroid may relieve itch. Fades spontaneously in 4–8wk.

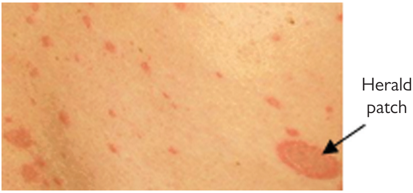

Figure 16.18 Pityriasis rosea showing herald patch

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Pityriasis (tinea) versicolor Chronic, often asymptomatic, fungal infection of the skin (*Pityrosporum orbiculare*). Common in humid/tropical conditions. In the UK often affects young adults and teenagers. On untanned, white skin appears as pinkish-brown, oval, or round patches with a fine superficial scale. In tanned or darker skin, patchy hypopigmentation occurs (Figure 16.19). Involves trunk ± proximal limbs.

Management Topical imidazole antifungal (e.g. clotrimazole cream bd), or topical selenium sulfide shampoo to all affected areas at night, washed off the following morning and repeated ×2 at weekly intervals. For resistant cases, try a systemic antifungal, e.g. itraconazole 200mg od for 1wk. Recurrences are common. Hypopigmentation may take some time to clear.

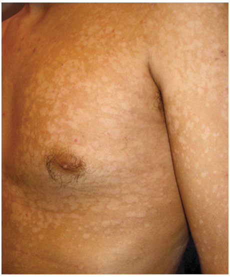

Figure 16.19 Pityriasis versicolor showing patchy hypopigmentation

Reproduced from Burge S. *et al*., *Oxford Handbook of Medical Dermatology*, second edition (2016) with permission from Oxford University Press.

Pityriasis alba Finely scaled white patches on face or arms (Figure 16.20). Affects children/young adults. Associated with atopy. Usually no treatment is required. Resolves spontaneously over months or years. If severe, refer to dermatology for confirmation of diagnosis. Treatment for severe cases is with topical steroids and/or PUVA.

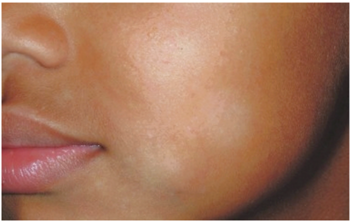

Figure 16.20 Pityriasis alba on a child's face

Reprinted from *Lookingbill and Marks' Principles of Dermatology*, sixth edition, Marks J.G. ***et al***., 184–95, Copyright © 2019, with permission from Elsevier.

Seborrhoeic keratosis (senile wart, basal cell papilloma) Common >60y. Often multiple—most commonly on trunk. Warty nodules, usually pigmented, 1–6cm diameter with a 'stuck-on' appearance (Figure 16.21). Pieces can be 'picked off'. *Cause*: unknown.

Features on dermoscopy ❗ Dermoscopy requires specialized training. Useful for distinguishing benign/malignant pigmented lesions. Typical dermoscopic features of seborrhoeic warts are:

- 'Fat fingers'
- Irregular crypts
- Fissures/ridges
- Blue-grey globules
- Light-brown fingerprint-like parallel structures
- Milia-like cysts—2 types: tiny, white starry and larger, yellowish cloudy

Management Reassurance. If removal is required, cryotherapy, curettage, shave biopsy, and excision biopsy are all effective.

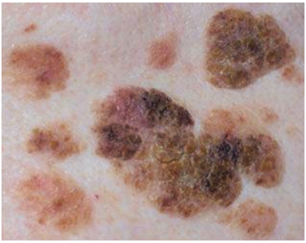

Figure 16.21 Seborrhoeic keratoses with typical 'stuck-on' appearance

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Further information

Primary Care Dermatology Society (2019) Pityriasis versicolor. 🖱 www.pcds.org.uk/clinical-guidance/pityriasis-versicolor

Sunlight and the skin

Skin conditions worsened by sunlight HSV (cold sores); lupus erythematosus (LE); porphyria; rosacea; Darier's disease.

Skin conditions improved by sunlight Acne; atopic eczema; pityriasis rosea; psoriasis (10% get worse).

Skin cancer ➔ p. 602 **Sunburn** ➔ p. 1097 **Pellagra** ➔ p. 615

Actinic (solar) keratosis Figure 16.22a. Single/multiple discrete scaly hyperkeratotic rough-surfaced areas over sun-exposed sites (e.g. dorsum of hands; head; neck). Occasionally occur on lower lip. More common with fairer skin types. May regress spontaneously or be pre-malignant.

Management Examine all the skin to look for skin cancer. Provide advice on UV protection (➔ p. 602). Treat individual lesions with fluorouracil cream od for 4wk, or cryotherapy. Treat field damage with diclofenac 3% gel, imiquimod cream, ingenol mebutate gel, or fluorouracil cream depending on size of affected area.

Complications Malignant change; cutaneous horn development (Figure 16.22b—treat with excision or curettage; send for histology).

Solar elastosis Sun-exposed skin is yellow, thickened, and wrinkled. No treatment. Advise to avoid sun exposure and wear sun block.

Solar urticaria Rare. Wheals appear within minutes of sun exposure. *Management*: sunscreens, avoidance. Refer to dermatologist if disabling.

Drug-induced photosensitivity Drugs may produce a light eruption in exposed areas by dose-dependent or allergic mechanisms. The reaction varies according to the drug. *Common examples are*: amiodarone, chlorpropamide, furosemide, griseofulvin, phenothiazines, sulfonamides, tetracyclines, thiazides, nalidixic acid, coal tar, plant-derived psoralens.

Plant-induced photodermatitis Contact dermatitis (➔ p. 581) in light-exposed areas resulting from local sensitization of the skin by contact with psoralens from plants (e.g. carrots, celery, fennel, parsnip, common rue, giant hogweed). Oils used in perfumes derived from plants (e.g. oil of bergamot) may also contain psoralens.

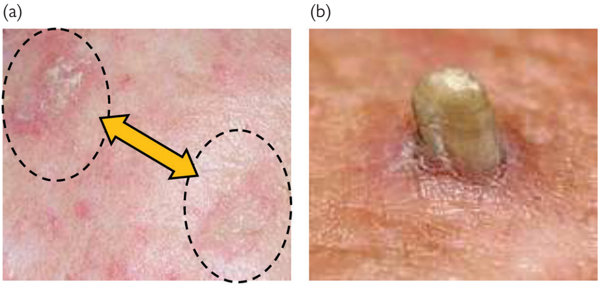

Figure 16.22 Actinic keratosis: (a) scalp lesions and (b) keratin horn

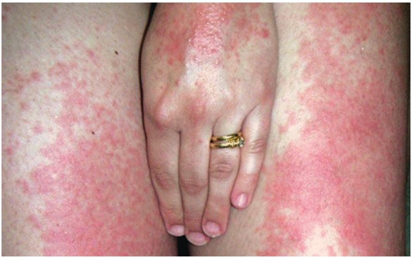

Figure 16.23 Polymorphic light eruption

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Polymorphic light eruption Pruritic papules, plaques ± vesicles appear in sun-exposed areas ~24h after exposure (Figure 16.23). Most common photodermatosis. ♀:♂ ≈2:1. *Cause*: unknown. *Management*: sunscreens and avoidance of sun exposure (sit in the shade, long sleeves, trousers, broad-brimmed hat); a short course of phototherapy in the Spring may help severe cases—refer to dermatology. Condition is non-scarring. *Differential diagnosis*: airborne contact dermatitis (plant or non-plant origin—plants are commonly from the Compositae family); drug-induced photosensitivity; lupus.

Actinic prurigo Rare. Starts in childhood. Papules and excoriations on sun-exposed sites. *Management*: sunscreens and avoidance. Refer to dermatology for confirmation of diagnosis and advice on further management.

Porphyria A group of rare, mostly inherited, metabolic disorders. Porphyrins are important in the manufacture of haemoglobin. Deficiency of enzymes in the porphyrin pathway results in build-up of intermediary metabolites which are toxic to the skin and nervous system. All require specialist management by either a general physician or a dermatologist. The main porphyrias are:

- Acute intermittent porphyria Intermittent attacks precipitated by many drugs. *Presentation*: fever, GI symptoms (vomiting, abdominal pain—can be severe); neuropsychiatric symptoms (hypotonia, paralysis, fits, impaired vision, peripheral neuritis, odd behaviour—even psychosis); *no* skin features. Urine may go deep red on standing
- Porphyria cutanea tarda Most common porphyria. Typically male alcoholics with liver damage. Presents with fragile blisters on the backs of hands which scar. *Management*: avoidance of alcohol and aggravating drugs (e.g. oestrogens); venesection; chloroquine
- Erythropoietic protoporphyria Autosomal dominant; starts in childhood; red blistering eruption leaving scars on hands and nose
- Variegate porphyria Autosomal dominant. Common in South Africa. Skin signs are like porphyria cutanea tarda, but abdominal pain and neuropsychiatric symptoms resemble acute intermittent porphyria

Further information

British Association of Dermatologists (2017) Actinic keratosis. 🖱 https://onlinelibrary.wiley.com/doi/full/10.1111/bjd.15107

Benign skin tumours

Seborrhoeic keratosis (senile wart, basal cell papilloma) ➔ p. 597

Chondrodermatitis nodularis ➔ p. 923 **Milia** ➔ p. 595

Naevus Benign proliferation of ≥1 normal constituent of the skin. The most common type is the melanocytic naevus or 'mole'. Most develop in childhood and adolescence. *Features*:

- Congenital Present at birth in 1% Caucasians (less in darker-skinned races). Usually >1cm diameter; if very large there is ↑ risk of malignancy
- Junctional Flat, round/oval, brown/black, 2–10mm diameter. Common sites: soles, palms, genitalia
- Intradermal Dome-shaped papule/nodule commonly on the face or neck. May be pigmented
- Compound <10mm diameter, smooth surface, variable pigmentation
- Blue Blue-coloured solitary naevus usually found on the face or extremities—especially hands and feet
- Halo Common in children/adolescents. Solitary/multiple. White halo of depigmentation surrounds naevus which then disappears. Associated with vitiligo. New onset in adults may suggest melanoma elsewhere

Differential diagnosis Freckle, lentigo, seborrhoeic keratosis, haemangioma (may be pigmented), dermatofibroma, pigmented BCC, melanoma.

Management Patients usually present if worried about a mole. Refer for urgent dermatology assessment if malignancy is suspected.

Skin tags Common. Small pedunculated polyps found in axillae, groin, neck, or on the eyelids. Reassure. Cosmetic removal can be achieved by snipping across the skin tag with scissors, cryotherapy, or diathermy.

Sebaceous cyst (epidermal cyst) Common. Round or oval, keratin-filled firm cysts, 1–3cm in diameter, within the skin. Usually a punctum is seen on the surface. Reassure. Treat any complicating bacterial infection with oral antibiotics (e.g. flucloxacillin 500mg qds). Excision is curative.

Dermatofibroma Common (♀ > ♂; young adult > elderly) and usually asymptomatic. Firm (sometimes pigmented) nodule 5–10mm in diameter that may occur following an insect bite or minor trauma (Figure 16.24). *Most common site*: lower legs. Treatment is not usually required. Refer urgently to dermatology if diagnostic uncertainty.

Keratoacanthoma Rapidly growing nodular tumour (<2cm in diameter) of sun-exposed skin of face/arms—Figure 16.25. A central keratin plug may fall out to leave a crater. Heals spontaneously over several months leaving a scar. *Differential diagnosis*: SCC. Lesions have a similar appearance to SCC and should be referred urgently to dermatology.

Campbell de Morgan spot (cherry angioma) Small bright red papules on the trunk in middle-aged/elderly patients. Usually require no treatment.

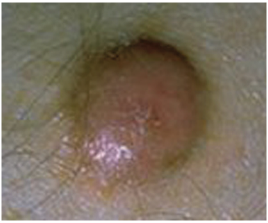

Figure 16.24 Dermatofibroma

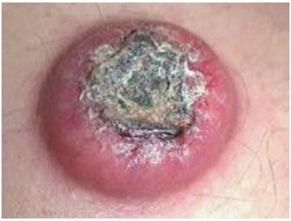

Figure 16.25 Keratoacanthoma

Keloid scar Proliferation of connective tissue presenting as firm smooth nodules/plaques in response to trauma. A scar is termed hypertrophic if changes are limited to the scar, but keloid if it extends beyond the limit of the original injury (Figure 16.26). *Most common sites*: upper back, chest, ear lobes. More common in dark-skinned races.

Management Consider steroid injection into the scar. Refer to dermatologist or plastic surgeon if this is ineffective—other treatments include cryotherapy, or topical silicone gel sheeting.

Pyogenic granuloma Bright red/blood crusted nodule that bleeds easily (Figure 16.27). Typically at the site of trauma (e.g. small cut) and enlarges rapidly over 2–3wk. Usually occurs in children/young adults. *Most common site*: finger. *Differential diagnosis*: melanoma.

Management

- Pregnant women May disappear spontaneously after delivery
- Other patients Curettage and cautery, or excision. Send for histology to exclude malignancy

Lipoma Common, benign tumour of fat. Presents as a soft mass in the subcutaneous tissue. Often multiple. Most common on trunk, neck, and upper extremities. Removal is rarely necessary. Refer for imaging/surgical excision if >5cm diameter, rapidly enlarging, and/or painful.

Patient information

British Association of Dermatologists Patient information leaflets on: seborrhoeic warts, keratoacanthoma, dermatofibroma, pyogenic granuloma. www.bad.org.uk

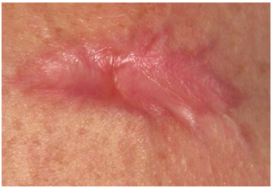

Figure 16.26 Keloid scar

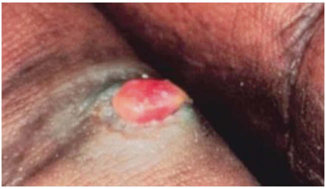

Figure 16.27 Pyogenic granuloma

Skin cancer

⚠ **Sun safety code** 80% of skin cancer is preventable.

- Take care not to burn—protect the skin with clothing including a shirt, trousers/skirt, hat, and ultraviolet-protective sun glasses
- Seek shade between 11 a.m. and 3 p.m.
- Apply high factor sunscreen (≥SPF 30 with high UVA protection)
- Take particular care to protect children in the sun

Cutaneous melanoma Every year in the UK there are 11,900 new cases of melanoma (lifetime incidence 1 in 60) and 2800 deaths. Incidence is rising. Particularly common in Caucasians. Frequently metastasizes and may present with metastases. *Types*:

- Superficial spreading 70% UK cases. ♀ > ♂. *Most common site*: lower leg in ♀ (50%); back in ♂. Macular lesion with variable pigmentation (Figure 16.28a)
- Nodular Figure 16.28b. 20% UK cases. ♂ > ♀. Most common on trunk. Firm nodule (pigmented, pink, or occasionally skin-coloured) grows rapidly and may ulcerate
- Lentigo maligna Arises in sun-damaged skin—usually on the face—and melanoma develops many years afterwards within it. Most common >60y—presents as an irregular brown/black patch
- Acral lentiginous Affects all races but accounts for 35–60% melanoma in black-skinned populations. Affects palms, soles, and nail beds. Often detected late

Risk factors UV exposure (especially sunburn when young, sun beds); genetic (10% have family history); multiple benign moles (>100 confers 10× ↑ risk); congenital naevus; previous melanoma; immunosuppression; fair skin type (red hair, blue eyes, and burns easily).

Assessment Encourage patients to report changes in moles early. Check the ABCDEF criteria:

- A Asymmetry of outline
- B Border irregularity
- C Colour variation
- D Diameter
- E Evolution—changes in size, shape, colour, and/or elevation
- F 'Funny looking' mole—'ugly duckling' moles that stand out from the others are very discriminatory for nodular melanoma

Management Use the 7-point checklist (Box 16.3) ± dermoscopy findings (if available) to identify changes needing referral. Refer all suspicious lesions for urgent dermatology assessment ± wide excision.

Dermoscopy ❗ Requires specialized training. Useful for distinguishing benign/malignant pigmented lesions.

Specialist treatment Best chance of cure comes with complete excision. Chemotherapy/radiotherapy are of little benefit but biological therapies (e.g. interferon, interleukin 2, ipilimumab, vemurafenib) are commonly used. A treatment vaccine is currently undergoing clinical trials.

Prognosis Relates to tumour depth at presentation. 5y survival: <0.76 mm deep—95%; >4mm deep—45%; metastases—10%.

Kaposi sarcoma ➔ p. 722

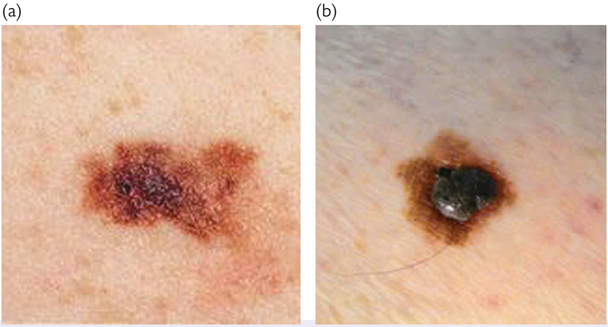

Figure 16.28 Malignant melanoma: (a) superficial spreading and (b) nodular

Box 16.3 The 7-point checklist for moles

Score 2 points for any major feature and 1 point for any minor feature. Lesions scoring ≥3 points are suspicious—refer.

Major signs

- Change in size—↑ in size
- Irregular colour
- Irregular shape—irregular border, asymmetry, elevation

Minor signs

- ≥7mm diameter
- Inflammation
- Oozing—including crusting/bleeding
- Change in sensation—including symptoms of minor irritation or itch

! One feature is enough to prompt referral if high level of suspicion. For low-suspicion lesions, monitor for change over 8wk.

Bowen's disease Intraepidermal SCC (Figure 16.29 ➔ p. 605). Common—typically occurs on the lower leg in elderly women. Lesions are flat-edged, pink/slightly pigmented scaly plaques (<5cm diameter) and may be solitary or multiple. *Risk factor*: chronic sun exposure, immunosuppression. 3–5% transform to SCC. Biopsy confirms diagnosis. Treatment is with fluorouracil cream od for 4wk, imiquimod cream, or cryotherapy (avoid lower legs). Excision is only usually required if diagnostic uncertainty.

Further information

NICE

- Suspected cancer: recognition and referral (2015, updated 2017). 🖱 www.nice.org.uk/guidance/ng12
- Melanoma: assessment and management (2015). 🖱 www.nice.org.uk/guidance/ng14

SIGN

- Cutaneous melanoma (2017). 🖱 www.sign.ac.uk/assets/sign146.pdf
- Management of primary cutaneous squamous cell carcinoma (2014). 🖱 www.sign.ac.uk/assets/sign140.pdf

Primary Care Dermatology Society

- Melanoma—an overview (2017). 🖱 www.pcds.org.uk/clinical-guidance/melanoma-an-overview1
- Bowen's disease (2017). 🖱 www.pcds.org.uk/clinical-guidance/bowens-disease

Squamous cell carcinoma (SCC) 20% of skin cancer in the UK. Most common >55y. ♂ > ♀. May metastasize (10%). Usually develops in sun-exposed sites, e.g. face, neck, hands. May start within an actinic (solar) keratosis (➔ p. 598) or *de novo* as a nodule which progresses to ulcerate and crust (Figure 16.30).

Causes Chronic sun damage, X-ray exposure, chronic ulceration and scarring (aggressive SCC may develop at the edge of chronic ulcers), smoking pipes and cigars (lip lesions), industrial carcinogens (tars, oils), wart virus, immunosuppression, genetic.

Management Refer urgently to dermatology. Treated with surgical excision ± LN biopsy. Large lesions may require skin grafting. Radiotherapy is an alternative for large lesions in elderly patients.

Basal cell carcinoma (rodent ulcer, BCC) Most common form of skin cancer—accounts for >75% of skin cancer in the UK. Locally invasive but rarely metastasizes. Tends to occur in middle-aged/elderly patients, may be multiple and affects any site. 3 major types (Figure 16.31—all can be pigmented):

- Nodular Most common. Often head or neck. Starts as small pearly nodule ± surface telangiectasia. May necrose centrally leaving a small crusted ulcer with pearly, rolled edge
- Superficial ≥1 pink patches with erosions and fine whipcord edge. Trunk is the most common site
- Morphoeic Waxy indurated plaque resembling a scar—usually on the face

Dermoscopy of BCC ❗ Requires specialized training:

- Focal ulceration
- Absence of pigment network
- Multiple blue-grey, round, or oval globules
- Linear and arborizing (tree branch-like) telangiectasia
- Structureless or leaf-like areas towards the edge of the lesion
- Spoke wheel areas (like spokes of a wheel radiating from central hub)

Causes Sun exposure, X-ray irradiation, chronic scarring, genetic predisposition, arsenic ingestion.

Management Complete excision is the ideal. Refer routinely—if low risk to GPwSI working in a community skin cancer clinic; if uncertain or any high-risk features (Box 16.4), to dermatology or plastic surgery.

Prognosis Recurrence rate is 5% at 5y for surgery, and slightly higher for non-surgical treatments. Development of new basal cell carcinoma at other sites is common.

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Primary Care Dermatology Society

- Squamous cell carcinoma (2017). 🖱 www.pcds.org.uk/clinical-guidance/squamous-cell-carcinoma
- Basal cell carcinoma—overview (2017). 🖱 www.pcds.org.uk/clinical-guidance/basal-cell-carcinoma-an-overview

SIGN (2014) Management of primary cutaneous squamous cell carcinoma. 🖱 www.sign.ac.uk/assets/sign140.pdf

Box 16.4 High-risk features of BCC

- Size >2cm
- Site—nose/paranasal folds, scalp/temples, lips
- Previously treated lesion
- Immunosuppression
- Genetic disorder associated with BCC, e.g. Gorlin's syndrome

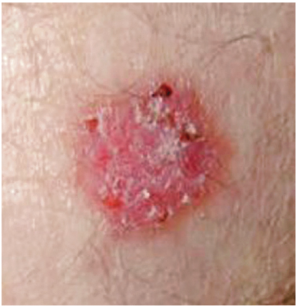

Figure 16.29 Bowen's disease

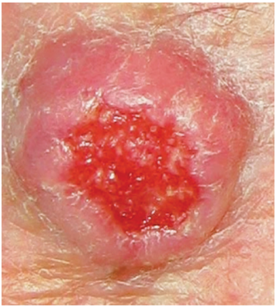

Figure 16.30 Squamous cell cancer showing ulcerated nodule

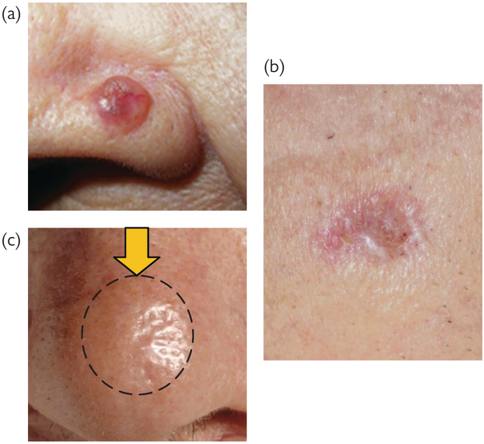

Figure 16.31 Basal cell cancer: (a) nodular BCC—pearly nodule showing surface telangiectasia, (b) superficial BCC—pink-red patch with focal erosions and subtle whipcord edge, and (c) morphoeic BCC (can be difficult to see)—waxy indurated plaque

Bacterial skin infection

Impetigo Superficial skin infection due to *Staph. aureus*. Very common in childhood. A thin-walled blister ruptures easily to leave a yellow crusted lesion. May occur anywhere but most common on the face. *Differential diagnosis*: HSV, fungal infection (e.g. ringworm). Lesions spread rapidly and are contagious. Avoid spreading to other children—no sharing of towels, face flannels, etc.; some schools/nurseries/childminders prohibit attendance until lesions are cleared. Reassure that it is non-scarring:

- Localized Treat with topical antibiotics (e.g. fusidic acid cream)
- Widespread Treat with oral flucloxacillin or clarithromycin.

Erysipelas and cellulitis Acute infection of the dermis. Often preceded by fever/'flu-like' symptoms. Usually affects face/lower leg. Appears as a unilateral, painful, tender, reddened area with a well-defined edge. Often the area is swollen and may blister. May be an obvious entry wound. *Differential diagnosis*: angio-oedema, contact dermatitis, gout.

Management

- Severe infection Admit for IV antibiotics
- If systemically well Mark the area before starting flucloxacillin 500mg qds or clarithromycin 500mg bd for 7–14d. Advise to seek help if infection is spreading or becoming systemically unwell
- Facial infection Treat with phenoxymethylpenicillin 500mg qds (flucloxacillin 500mg qds if *Staphylococcal* infection suspected; clarithromycin 500mg bd if penicillin allergic). Have a low threshold for admission
- Recurrent infections (>2 episodes at one site) may need prophylactic long-term penicillin (e.g. phenoxymethylpenicillin 250mg od or bd) with attention to skin care and management of any lymphoedema

Boils and carbuncles

- Boil (furuncle) Acute infection of a hair follicle, usually with *Staph. aureus*. A hard, tender, red nodule surrounding a hair follicle becomes larger and fluctuant after several days. Occasionally associated with fever ± malaise. Later may discharge pus and a central 'core' before healing; may leave a scar. *Predisposing factors*: usually absent—DM; HIV; obesity; blood dyscrasias; immunosuppressive drugs
- Carbuncle Swollen, painful area discharging pus from several points. Occurs when a group of hair follicles become deeply infected, usually with *Staph. aureus*. May be associated with fever ± malaise. *Predisposing factors*: malnutrition, cardiac failure, drug addiction, severe generalized dermatosis, prolonged steroid therapy, DM

Management

- Non-fluctuant lesions Apply moist heat to relieve discomfort, help localize the infection, and promote drainage
- If fever/surrounding cellulitis or lesion is on the face Treat with oral antibiotics, e.g. flucloxacillin 500mg qds for 7d—clarithromycin 500mg bd is an alternative if allergic to penicillin
- If large, but localized, painful, and fluctuant Consider incision and drainage (❗ do not attempt if you are not confident). Admission may be needed if young or uncooperative child, or the boil in a sensitive

area, e.g. genital region, face, neck, axilla, breast. Afterwards treat with oral antibiotics until inflammation resolves

- Admit If not settling with primary care treatment
- If recurrent or chronic Take swabs for culture from lesions and carrier sites (nose, axilla, and groin); treat carrier sites with topical antibiotic (e.g. Naseptin® qds for 10d). Advise improved hygiene and use of antiseptics in the bath (e.g. chlorhexidine); consider long-term antibiotics (e.g. clarithromycin 500mg od)

Folliculitis Superficial infection of the hair follicles usually caused by *Staph. aureus*. Presents as pustules in hair bearing areas, e.g. legs, beard area. *Risk factors*: obesity, DM, occlusion from clothing, topical steroid use. *Differential diagnosis: Malassezia* folliculitis (➔ p. 583).

Management Exclude DM; treat with topical antiseptic, or if resistant topical or systemic antibiotics (e.g. fusidic acid cream or oral flucloxacillin). *If recurrent or chronic*: Treat as for recurrent boils.

Acute paronychia Infection of the skin and soft tissue of the proximal and lateral nail fold, most commonly caused by *Staph. aureus*. Often originates from a break in the skin or cuticle as a result of minor trauma, e.g. nail biting. Skin and soft tissue of the proximal and lateral nail fold are red, hot, and tender; nail may appear discoloured/distorted. Treat in the same way as for a boil.

Staphylococcal whitlow (felon) Infection involving the bulbous distal pulp of the finger following trauma or extension from an acute paronychia. The finger bulb is red, hot, oedematous, and usually exquisitely tender. Onset of pain is rapid and there is swelling of the entire finger pulp. *Differential diagnosis*: herpetic whitlow—➔ p. 609.

Management

- If fluctuant Admit for drainage and antibiotics
- If non-fluctuant Elevate, apply moist heat (e.g. soak in hot water), and treat with oral antibiotics; if this fails, admit for incision and drainage

Wound infection Suspect if a wound becomes painful. Look for swelling, erythema, wound tenderness ± pus. *Risk factors*:

- Malnutrition
- DM
- Carcinomatosis
- Steroid therapy
- Infection near the site of incision
- Contamination of the wound

Management If pus is present send a swab for M,C&S:

- If indurated + infection is localized to the wound suspect *Staphylococcus*. Treat with flucloxacillin 500mg qds or clarithromycin 500mg bd
- If cellulitis around the wound suspect *Streptococcus*. Treat with phenoxymethylpenicillin 500mg qds or clarithromycin 500mg bd
- If foul smell, suspect anaerobes—treat with metronidazole 400mg tds

Give adequate analgesia; dress the wound frequently; review regularly; allow pus to drain. If a surgical wound, refer back to the operating surgeon if simple measures are ineffective.

⚠ **Necrotizing fasciitis** Life-threatening soft tissue infection. Usually occurs in otherwise healthy individuals after surgery/trauma (often minor). Ill-defined erythema + high fever. The wound rapidly becomes necrotic. Admit as an emergency for IV antibiotics ± surgery.

Viral skin infection

Systemic viral infections ➔ p. 628 **HIV infection** ➔ p. 720

Viral warts Common and benign. Due to infection of epidermal cells with human papilloma virus (HPV). >50 types identified. The virus is transmitted by direct contact. Immunosuppressed patients are particularly vulnerable.

Genital warts ➔ p. 724

Common warts Dome shaped papules with papilliferous surface. Usually >1. Most common on hands but may affect other areas. In children 30–50% disappear spontaneously in <6mo.

Plantar warts (verrucas) on soles of feet. Common in children. Pressure makes them grow into the dermis. Often painful. Characterized by dark punctate spots on the surface (may need to pare callus off to see). Warts group together to form mosaics.

Plane warts Smooth, flat-topped papules often slightly brown in colour. Most common on face/backs of hands. Usually >1. Manage as for common/plantar warts. Eventually resolve spontaneously. May show Koebner phenomenon.

Treatment of common, plantar, and plane warts Refer immunosuppressed patients for specialist advice. Otherwise treatment is usually unnecessary. If patients are insistent, advise OTC topical salicylic acid preparations, e.g. Duofilm®, Salactol®.

HPV vaccination ➔ p. 725

Herpes simplex infection HSV is transmitted by direct contact with lesions. Lesions may appear anywhere on the skin or mucosa but are most frequent around the mouth and on the lips, conjunctiva, cornea, and genitalia. Diagnosis is usually clinical.

Primary HSV stomatitis After a prodromal period (<6h) of tingling, discomfort, or itching, small tense vesicles appear on an erythematous base. These burst to form multiple, small, painful mouth ulcers. Infection may be accompanied by systemic symptoms, e.g. fever, malaise and tender lymph nodes. ❗ May be asymptomatic and go unnoticed.

Management of 1° HSV stomatitis Give symptomatic relief—try analgesic mouthwashes, e.g. benzydamine; healing occurs in 8–12d. If seen <48h after onset prescribe oral antivirals, e.g. aciclovir 200mg 5×/d. If unable to take fluids/becoming dehydrated, admit for IV fluids.

Recurrent infection (cold sores) Figure 16.32. After initial infection, HSV remains dormant in the nerve ganglia. Recurrent eruptions can occur, precipitated by overexposure to sunlight, febrile illnesses, physical or emotional stress, or immunosuppression. The trigger stimulus is often unknown. Recurrent disease is generally less severe and more localized. Treat with aciclovir cream 5% ×5/d if needed (available OTC).

Genital herpes ➔ p. 724

Neonatal herpes ➔ p. 724

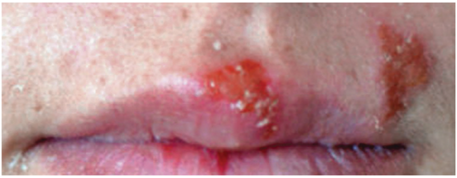

Figure 16.32 Cold sores

Herpetic whitlow Swollen, painful, and erythematous lesion of the distal phalanx with multiple vesicles visible. Results from inoculation of HSV through a skin break or abrasion. Treat with topical/oral aciclovir.

Molluscum contagiosum DNA pox virus infection spread by contact. Common in pre-school children. Less common in adults: associated with contact sports (e.g. wrestling), or sexually transmitted.

Childhood molluscum Presents as discrete pearly pink, umbilicated papules 1–3mm diameter (Figure 16.33). If squeezed, papules release a cheesy material. Lesions are multiple and grouped—usually on the trunk, face, or neck. Untreated lesions resolve spontaneously after 12–18mo. Treatment is not needed. Seek specialist advice if immunocompromised.

Genital molluscum Presents similarly to childhood molluscum but usually confined to the genital area. No treatment is needed but screen for other sexually transmitted infection

Orf Solitary, red, rapidly growing papule <1cm diameter—often on a hand. Evolves into a painful, purple pustule (Figure 16.34). There is usually a history of close contact with sheep (e.g. vet, farmer), or cows (when termed *milker's nodule*). *Cause*: parapox virus. *Incubation*: ~6d. Resolves spontaneously in 2–4wk. Complications include 2° infection (treat with topical/systemic antibiotics); erythema multiforme; lymphangitis.

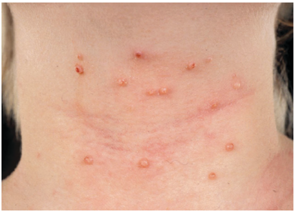

Figure 16.33 Molluscum contagiosum

Reproduced from Lewis-Jones S, *Paediatric Dermatology* (2010), with permission from Oxford University Press.

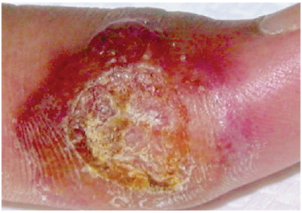

Figure 16.34 Orf on a thumb

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Fungal infection

Table 16.10 Presentation of candidiasis

Presentation	Symptoms	Differential diagnosis
Genital infection 'Thrush' ➔ p. 713	♀ >> ♂. Itchy, sore vulvovaginitis ± white plaques on mucous membranes and cheesy discharge. Men develop a similar clinical picture	Psoriasis; lichen planus; lichen sclerosus; other causes of vaginal discharge
Intertrigo	Reddened, moist, glazed area in the submammary, inguinal, or axillary folds. In wet workers may occur between digits. Patients may present with skin changes and/or itch	Psoriasis; tinea cruris; seborrhoeic dermatitis; bacterial skin infection
Oral	Sore mouth; poor feeding in infants. Most common in babies, patients with poor oral hygiene, or the elderly with false teeth. White plaques visible on buccal mucosa which can be wiped off ± angular stomatitis	Lichen planus; epithelial dysplasia
Nappy candidiasis	Babies—in the nappy area	Nappy rash—➔ p. 882
Chronic paronychia	Often seen in wet workers. Presents with chronic nail fold inflammation	Bacterial infection; chronic eczema
Systemic candidiasis	Occurs in immunosuppressed individuals (e.g. HIV, malignancy). Red nodules may appear on the skin	

There are two major groups of fungal skin infections seen in the UK.

Candidiasis Uniform commensal of the mouth/GI tract which causes opportunistic infection. *Risk factors*: moist, opposing skin folds; obesity; DM; neonates; pregnancy; poor hygiene; humid environment; wet work occupation; use of broad-spectrum antibiotic. Presentation—Table 16.10.

Dermatophyte infection Tinea denotes fungal infection. Common. Affects skin, hair or nails. Skin scrapings or nail clippings may confirm diagnosis. Presentation—Table 16.11.

General measures for prevention of fungal infections Keep body folds separated and dry (e.g. with dusting powder) and minimize hot and humid conditions (e.g. advise open footwear).

Topical treatment of fungal infections

- **Mouth lesions** Remove tongue deposits with a toothbrush by brushing 2×/d. Treat with oral suspensions or gels (e.g. nystatin, miconazole). If false teeth, place imidazole gel on the teeth before insertion and sterilize overnight with dilute hypochlorite solution (e.g. Milton®)
- **Genital lesions** Treat with imidazole cream or pessaries
- **Nail infections** If confined to 1 or 2 nails, use a lacquer or paint (e.g. amorolfine). Apply 1–2×/wk after filing/cleansing for 6mo (fingernails) or 9–12mo (toenails). Avoid nail varnish/artificial nails while treating. If more extensive or ineffective, consider oral therapy
- **Skin lesions** Use imidazole cream, spray, or powder; or terbinafine cream

Systemic treatment Use for resistant, recurrent, extensive, or systemic infection, and nail or scalp infection. Warn about side effects.

- **Oral, mucocutaneous, or systemic candidiasis** Oral fluconazole 50mg od for 1–2wk—higher doses/prolonged therapy may be needed if immunosuppressed (seek specialist advice)
- **Genital candidiasis** Single oral dose of 150–200mg fluconazole
- **Tinea pedis/manuum** Oral terbinafine (250mg od for 2–6wk) or itraconazole (100mg od for 30d or 200mg bd for 7d)
- **Tinea cruris/corporis** Oral terbinafine (250mg od for 2–4wk for tinea cruris and 4wk in tinea corporis) or itraconazole (100mg od for 15d or 200mg od for 7d)
- **Nail infection** Consider if topical treatment is unsuccessful or >2 nails involved. Confirm the diagnosis with nail clipping mycology before treatment with oral terbinafine (250mg od for 6wk–3mo) or pulsed itraconazole (200mg bd for 7d repeated after 21d ×2 for fingernail infections and ×3 for toenail infections)
- **Scalp infection** If kerion (pustular boggy mass) is suspected, refer to dermatology. Otherwise, oral terbinafine (250mg od) or griseofulvin (500mg–1g od) depending on sensitivities. ⚠ Griseofulvin is teratogenic—advise ♀ to avoid pregnancy during treatment and for 1mo afterwards and ♂ to use contraception during treatment and for 6mo afterwards

Pityriasis versicolor ➔ p. 596

Table 16.11 Dermatophyte infections

Tinea	Affects	Presentation	Differential diagnosis
Corporis 'Ringworm'	Trunk or limbs	Single/multiple plaques with scaling and erythema especially at the edges. Lesions enlarge slowly and clear centrally (hence 'ringworm')	Discoid eczema Psoriasis Pityriasis rosea
Cruris 'Jock itch'	Groin ♂ > ♀ Common in athletes	Associated with tinea pedis. Involves upper thigh (+ scrotum rarely). Red plaque with scaling especially at the edge	Intertrigo Candidiasis Erythrasma
Pedis 'Athlete's foot'	Feet ♂ > ♀ Young > old	Itchy, maceration between toes. *Risk factors*: swimming; occlusive footwear; hot weather	Contact dermatitis Psoriasis Pompholyx
Capitis	Hair and scalp	Defined, inflamed scaly areas ± alopecia with broken hair shafts	Alopecia areata Psoriasis Seborrhoeic eczema
Unguium	Nails—prevalence ↑ with age; rare in children Toenails > fingernails	Begins at distal nail edge; progresses proximally to involve the whole nail. Eventually results in thickening, yellowing, and crumbling of the nail plate. Tinea pedis often coexists	Psoriasis Trauma Candidiasis

Infestations

Head lice Most common in children age 4–11y (♀ > ♂) but may occur in anyone. Contrary to popular belief, lice infest clean as often as dirty hair. Adult lice are about the size of a sesame seed, brownish-grey in colour, and wiggle their legs (Figure 16.35). Only adults are contagious. Spread by close head-to-head contact. Lice do not jump/fly and do not remain viable away from a host.

Symptoms/signs Normally asymptomatic. Detected by contact tracing of other cases or routine inspection at home or school. Occasionally present as itchy scalp. Presence of 'nits' (egg shells—white dots attached to hair), eggs, or dead lice indicate past infection—a moving louse must be found to confirm active infection.

Detection After washing hair, apply conditioner and comb with fine-tooth detector comb (available from pharmacy). In at-risk groups (e.g. schoolchildren) repeat weekly. Lice are removed by the comb and seen trapped in its teeth.

Management Treat all household contacts simultaneously.

- Prophylactic preparations No evidence of effectiveness
- Dimeticone Lotion or spray. Coats lice and interferes with their water balance by preventing the excretion of water. Advise to rub into dry hair and scalp in the evening, allow to dry naturally, then shampoo off the next morning. Repeat after 7d
- Insecticides Effective. 4 types: malathion, phenothrin, permethrin (all available OTC but NHS prescriptions are often sought), and carbaryl (prescription only). Malathion and phenothrin/permethrin are used as 1st/2nd line; carbaryl is reserved for 3rd line. Apply according to the manufacturer's instructions using 2 applications, 7d apart. Check wet, conditioned hair with a detector comb before the 1st application then every 2d until 2–3d after the 2nd application. Supply enough for 2 applications. Shampoos are ineffective—use lotions, liquids, or cream rinses
- Mechanical clearance Wet-comb conditioned hair with a fine-tooth comb until all lice are removed and repeat at 3–4d intervals for 2wk. Alternative to insecticides but requires motivation
- Other methods of treatment e.g. electric combs, aromatherapy (tea tree oil), herbal treatments—no evidence supporting use

❗ If pregnant/breastfeeding, treat with wet combing or dimeticone

Contact tracing All cases—trace close contacts over the past month and ask them to check their scalps for lice/treat as needed.

Reinfestation/resistance to treatment 3 possible reasons:

- Reinfestation Lice found are large adults only. Ask patient to check close contacts again. Re-treat with a different insecticide
- Incorrect use of insecticide/mechanical clearance Lice are at mixed stages of development. Check procedure and make sure instructions are understood. Repeat treatment with a different insecticide
- Resistance to insecticide Lice are seen at all stages of development. Re-treat with another product

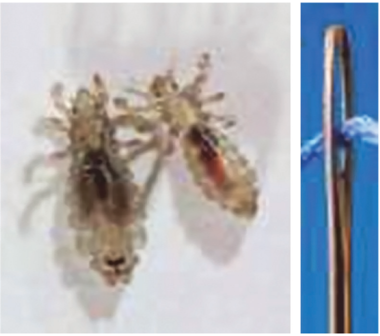

Figure 16.35 Head lice with needle and thread to give an idea of size

Crab (pubic) lice ➔ p. 725

Scabies Extremely contagious. The scabies mite (*Sarcoptes scabei*) is ~0.5 mm long and spread by direct physical contact. Average infection consists of 12 mites.

Presentation Symptoms of intense itching appear 4–6wk after infection. Examination reveals burrows (irregular, tortuous, and slightly scaly <1cm long) on the sides of fingers, wrists, ankles, and nipples. May form rubbery nodules on genitalia. Itching results in excoriations. Mites may be visible with dermoscopy. Untreated infection becomes chronic.

Differential diagnosis Lichen planus; dermatitis herpetiformis; papular urticaria; eczema.

Management Treat with scabicide, e.g. permethrin 5% or malathion lotion. Apply according to manufacturer's instructions. All close contacts need treatment simultaneously, which may result in all occupants of a residential home being treated. Apply to whole body including scalp, neck, face and ears. Ensure finger/toe webs are covered and brush lotion under the ends of fingernails/toenails. Reapply to whole body after 1wk and to hands alone if washed with soap <8h after application. Advise patients to launder all worn clothing and bedding after application. Itching may persist for some time after elimination of infection—use chilled crotamiton lotion and/or sedating oral antihistamines for symptomatic relief.

Complications 2° infection (treat with topical or systemic antibiotics).

Crusted 'Norwegian' scabies Affects debilitated or immunosuppressed patients There is overwhelming infection with >10,000 mites. Typically the infestation is not itchy but presents with a crusted skin rash often misdiagnosed as psoriasis. Under the microscope, crusts are seen to contain hundreds of scabies mites. Treatment is as for scabies; resistant cases can be treated with ivermectin which is available only on a named patient basis in the UK—discuss with a specialist dermatologist. ❗ People in contact with sufferers may develop a red, itchy rash themselves—treat with insecticide as for scabies.

Skin changes associated with internal conditions

Table 16.12 Systemic conditions associated with skin changes

Condition	Associated skin changes
Addison's disease	Pigmentation, vitiligo
Cushing's disease	Pigmentation, hirsutism, striae, acne, truncal obesity, moon facies, buffalo hump
Diabetes mellitus	• Diabetic dermopathy Depressed pigmented scars on the shins • Necrobiosis lipoidica Shiny, atrophic yellowish-red plaques on the shins. Affects <1% diabetics but limited to those with DM or who will later develop DM • Granuloma annulare Palpable annular lesions on hands, feet, or face. Only rarely associated with DM. Fades spontaneously in <12mo. Differentiate from ringworm • Xanthoma (see 'Hyperlipidaemia') • Fungal infection (➲ p. 610) • Neuropathic ulcers (➲ p. 584)
Drug eruptions	Common. Withdrawal of the offending drug usually results in clearance of the eruption in <2wk. Simple emollients ± topical steroids may ease symptoms in the interim. Occasionally patients with severe reactions may require admission for supportive treatment until effects of the drug wear off. Stevens–Johnson syndrome (erythema multiforme—➲ p. 569)
Hyperlipidaemia	Xanthoma Yellowish lipid deposits in the skin—may be eruptive (like a rash), tendinous, plane (palmar creases), tuberous (knees, elbows) Xanthelasma Yellowish plaques on eyelids. Not always associated with hyperlipidaemia
Inflammatory bowel disease	Crohn's disease Perianal abscess, sinuses, or fistulae; erythema nodosum, Sweet's disease (dark red plaques on face, arms, and legs), clubbing Ulcerative colitis Pyoderma gangrenosum, erythema nodosum, Sweet's disease (see 'Crohn's disease'), clubbing
Liver disease	Pruritus, spider naevi, erythema, white nails, pigmentation, xanthomas (see 'Hyperlipidaemia')
Malabsorption	Dry itchy skin, ichthyosis, eczema, oedema Dermatitis herpetiformis (associated with coeliac disease)

(*Continued*)

Table 16.12 *(Contd.)*

Condition	Associated skin changes
Malignancy	Acanthosis nigricans Rare, epidermal thickening and pigmentation in flexures and neck. Associated with GI malignancy Mycosis fungoides Lymphoma that evolves in the skin. Slowly progressive becoming systemic only in terminal stages. May resemble psoriasis or eczema in early stages Paget's disease of the nipple ➔ p. 674 Skin secondaries Most commonly breast, GI, ovary, lung, or haematological Lymphoedema ➔ p. 1025
Other conditions occasionally associated with malignancy Flushing, generalized pruritus, hyperpigmentation, ichthyosis, dermatomyositis, erythroderma, hypertrichosis, pyoderma gangrenosum, superficial thrombophlebitis, tylosis	
Malnutrition	Iron deficiency Alopecia, koilonychia, itching Scurvy Vitamin C deficiency—bleeding gums, woody oedema, perifollicular oedema Protein deficiency Pigmentation, dry skin, oedema, pale brown/orange hair Pellagra Nicotinic acid deficiency—photosensitive dermatitis in sun exposed areas ± dementia/diarrhoea
Neurofibromatosis	➔ p. 552
Pregnancy	Pigmentation, spider naevi, abdominal striae, pruritus, pruritic urticarial papules and plaques of pregnancy (PUPP—1 in 240 pregnancies), pemphigoid gestationis (rare)
Sarcoidosis	Nodules, plaques, erythema nodosum, dactylitis, lupus pernio (dusky-red infiltrated plaques on nose ± fingers)
Thyroid disease	Hypothyroidism Alopecia, coarse hair, dry, puffy brownish-yellow skin Thyrotoxicosis Pink, soft skin, hyperhidrosis, alopecia, pigmentation, onycholysis, clubbing, pretibial myxoedema (raised erythematous plaques on shins—topical steroids may help)
Tuberous sclerosis	Adenoma sebaceum Red/yellow fibromatous plaques—usually around nose Periungual fibroma Pink, fibrous projections under nail folds Ash-leaf macules White, oval macules—best seen under Wood's light (if available) Shagreen patches Yellowish naevi with cobblestone surface—found on the back

Infectious diseases covered in other chapters

Infection	Page	Infection	Page
Aspergillosis	➜ p. 299	Influenza	➜ p. 292
Bacterial vaginosis	➜ p. 713	Kawasaki's disease	➜ p. 501
Boils and carbuncles	➜ p. 606	Listeriosis	➜ p. 798
Brain abscess	➜ p. 532	Lyme disease	➜ p. 571
Bronchiolitis	➜ p. 855	Meningitis	➜ p. 1056
Bronchitis (acute)	➜ p. 292	Molluscum contagiosum	➜ p. 609
Campylobacter	➜ p. 348	Mycoplasma	➜ p. 298
Candidiasis—genital	➜ p. 713	Norovirus	➜ p. 348
Candidiasis—mouth	➜ p. 610	Osteomyelitis	➜ p. 478
Candidiasis—skin	➜ p. 610	Otitis externa	➜ p. 922
Cellulitis and erysipelas	➜ p. 606	Otitis media	➜ p. 924
Chlamydia—genital	➜ p. 716	Pneumocystis	➜ p. 299
Chlamydia—neonatal	➜ p. 716	Pneumonia—adult	➜ p. 294
Chlamydia—respiratory	➜ p. 298	Pneumonia—childhood	➜ p. 854
Cholecystitis	➜ p. 401	Polio	➜ p. 551
CJD	➜ p. 543	Prostatitis	➜ p. 430
Common cold	➜ p. 292	Rotavirus	➜ p. 349
Conjunctivitis	➜ p. 944	Salmonella	➜ p. 348
COVID-19	➜ p. 299	SARS	➜ p. 299
Croup	➜ p. 915	Scabies	➜ p. 613
Cryptosporidium	➜ p. 348	Septic arthritis	➜ p. 494
E. coli diarrhoea	➜ p. 348	Sinusitis	➜ p. 920
Encephalitis	➜ p. 1056	Skin infection—bacterial	➜ p. 606
Endocarditis (infective)	➜ p. 244	Skin infection—fungal	➜ p. 610
Epiglottitis	➜ p. 915	Skin infection—viral	➜ p. 608
Gastroenteritis	➜ p. 348	Syphilis	➜ p. 725
Glandular fever	➜ p. 913	Tonsillitis	➜ p. 912
Gonorrhoea	➜ p. 717	Toxoplasmosis	➜ p. 798
Guillain–Barré	➜ p. 516	*Trichomonas vaginalis*	➜ p. 716
Head lice	➜ p. 612	Tuberculosis (TB)	➜ p. 296
Helicobacter pylori	➜ p. 354	URTI (childhood viral)	➜ p. 854
Hepatitis A/E	➜ p. 394	UTI in adults	➜ p. 422
Hepatitis B/C	➜ p. 718	UTI in childhood	➜ p. 856
Herpes simplex—eye	➜ p. 946–7	UTI in pregnancy	➜ p. 798
Herpes simplex—genital	➜ p. 724	Warts—genital	➜ p. 724
Herpes simplex—skin	➜ p. 608	Warts—skin	➜ p. 608
HIV	➜ p. 722	Whooping cough	➜ p. 298
Impetigo	➜ p. 606		

Chapter 17

Infectious disease

Immunization *618*
Symptoms, signs, and notification of infectious disease *620*
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Streptococcal and staphylococcal infections *630*
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Immunization

Immunity can be induced in 2 ways

- **Active immunity** Induced using inactivated or attenuated live organisms or their products. Act by inducing cell-mediated immunity and serum antibodies. Generally long-lasting
- **Passive immunity** Results from injection of human immunoglobulin. The protection afforded is immediate but lasts only a few weeks

Storage of vaccines Follow manufacturers' instructions. Do not store vaccines in the door of a vaccine fridge and make sure there is a maximum and minimum thermometer in the fridge. Record readings regularly and discard vaccines if not stored at the correct temperature.

Administration of vaccines Only suitably trained GPs/nurses should give immunizations. Check immunization is needed and the patient is fit. Check consent has been obtained and that immunizations are the correct ones and in date. Ensure resuscitation facilities are available. Record vaccine expiry date/batch number. Reconstitute vaccine (if necessary) and give according to manufacturer's instructions. Record date and site in the medical notes.

Childhood immunization In the UK, routine vaccinations for the under 5s are usually done in the GP surgery. Routine vaccinations for older children are normally done through the school health service. Schedule for childhood immunizations—Table 17.1

Adult immunization

Influenza and pneumococcal vaccination Available as a directed enhanced service—existing practices do not have preferred provider status. Additional payments are available through the Quality and Outcomes Framework for ensuring at-risk patients receive vaccination.

Other necessary vaccinations Can be provided as an additional service. Opting out incurs a 2% ↓ in global sum. A list of eligible vaccinations and terms of eligibility is available on the BMA website (☞ www.bma.org.uk). Includes hepatitis A, cholera, and typhoid vaccinations; travel vaccinations that do not fall into these criteria can be administered as a private service.

Contraindications to vaccination For specific contraindications to individual vaccinations consult the 'Green Book'. General rules:

- **Acute illness** Delay until fully recovered. Minor ailments without fever or systemic upset are not reasons to postpone immunization
- **Severe local reaction to previous dose** Extensive area of redness/swelling that involves much of the antero-lateral surface of the thigh or a major part of the circumference of the upper arm
- **Severe generalized reaction to a previous dose** Fever ≥39.5°C <48h after vaccination; anaphylaxis, bronchospasm, laryngeal oedema, and/or generalized collapse; prolonged unresponsiveness; prolonged high-pitched or inconsolable screaming for >4h; convulsions or encephalopathy <72h after vaccination

Table 17.1 UK schedule of childhood immunization

Disease	Vaccine	Age
Tuberculosis (BCG)	BCG	High-risk neonates
Diphtheria, tetanus, pertussis, polio, *Haemophilus influenza* type b (Hib), and hepatitis B	DTaP/IPV/Hib/HepB	2, 3, and 4mo
Pneumococcal (13 serotypes)	Pneumococcal conjugate vaccine (PCV)	2, 4, and 12mo
Rotavirus vaccine (oral)	Rotavirus (oral)	2 and 3mo
Meningococcus type B	MenB (left thigh)	2, 4, and 12mo
Meningococcus type C, *Haemophilus influenzae* type b	Hib/MenC	12mo
Measles, mumps, rubella	MMR	12mo and 3y 4mo
Influenza (➔ p. 293)	≥2–17y: Live attenuated influenza vaccina (LaIV) (intranasal—both nostrils) <2y: Quadrivalent inactivated influenza vaccine (IM)	Annually—high-risk children + all children aged 2–8y*
Diphtheria, tetanus, pertussis, polio	DTaP/IPV	3y 4mo
HPV vaccination	HPV (2 doses 6–24mo apart)	12–13y
Tetanus, diphtheria, polio	Td/IPV	14y
Meningococcus A, C, W, and Y	MenACWY	14y

*Over coming years will be extended to age 2–17y

Contraindications to live vaccines BCG; measles; mumps; oral typhoid; rubella; yellow fever. *Do not* give live vaccines:

- To pregnant women or immunocompromised patients—those on high-dose steroids for >1wk (>1mg/kg/d prednisolone for children or ≥40mg/d for adults); if haematological malignancy; if radiotherapy/chemotherapy within 6mo; or another immunodeficiency syndrome
- <3wk after another live vaccine (but 2 live vaccines may be given together at different sites), or
- With immunoglobulin (from 3wk before to 3mo after)

❗ Patients with HIV who are not severely immunosuppressed may have live vaccines except BCG and yellow fever.

Vaccine damage payments Only payable if a patient is >60% impaired by a vaccination given within the NHS. Recipients receive a lump sum. Further information: ☎ 01772 899944 🖱 www.gov.uk.

Further information

Public Health England (2014) The Green Book: Immunisation against infectious disease. 🖱 www.gov.uk/government/collections/immunisation-against-infectious-disease-the-green-book

Public Health England (2018) Complete routine immunisation schedule. 🖱 www.gov.uk/government/publications/the-complete-routine-immunisation-schedule

Symptoms, signs, and notification of infectious disease

Specific symptoms and signs of infection depend on the infecting organism and organs affected. For example, a chest infection will cause respiratory symptoms; a urine infection urinary tract symptoms. Symptoms suggesting an infectious cause include:

Lymphadenopathy Palpable enlargement of the LNs:

Benign causes

- Infective Bacterial—pyogenic, TB, *Brucella*; fungal; viral—EBV, CMV, HIV; toxoplasmosis; syphilis
- Non-infective Sarcoid, connective tissue disease (rheumatoid arthritis); skin disease (eczema, psoriasis); drugs (phenytoin); berylliosis

Malignant causes Lymphoma, CLL, ALL, metastases.

Management in adults Refer immediately for urgent investigation if:

- Rapidly growing
- Non-tender, firm/hard lymph node >3cm diameter
- LNs associated with other unexplained signs of ill health (night sweats, weight loss, persistent fever)
- LNs associated with other sinister signs, e.g. petechial rash (same day assessment), suspected head or neck tumour
- Enlarged supraclavicular nodes in the absence of local infection

Most enlarged LNs are reactive LNs—suggested by a short history; soft, tender, mobile lump; and concurrent infection. If there are no sinister features, give these 2wk to settle. If not settling, check FBC, ESR, ± EBV screen. Refer lymphadenopathy >1cm diameter persisting for >6wk for urgent further investigation.

Management in children Refer to paediatrics urgently, particularly if there is no evidence of local infection, if ≥1 of:

- Non-tender, firm/hard LN
- LN >2cm diameter
- Progressively enlarging LNs
- LNs associated with other signs of ill health (e.g. fever, weight loss)
- Enlarged axillary LNs in the absence of local infection or dermatitis
- Supraclavicular node involvement

Investigate with FBC and blood film if generalized lymphadenopathy.

Table 17.2 Normal temperature as measured in different locations

Place of measurement	Normal range
Oral	35.5–37.5°C (95.9–99.5°F)
Rectal	36.6–38.0°C (97.9–100.4°F)
Axillary	34.7–37.3°C (94.5–99.1°F)
Ear	35.8–38.0°C (96.4–100.4°F)

Pyrexia/fever Oral temperature raised above 37.5°C. Normal range varies according to where measured—Table 17.2. *Common causes*:

Infection By far the most common cause in general practice:

- Viral infection (e.g. HIV, EBV, URTI, influenza)
- UTI
- Chest infection
- Tonsillitis
- Otitis media
- Sinusitis
- Cholecystitis
- Cellulitis

❗ Do not forget tropical diseases, e.g. malaria in patients returning from abroad. Think of TB and SBE—especially in high-risk patients.

Cancer Lymphoma; leukaemia; solid tumours (e.g. hypernephroma).

Immunogenic causes Connective tissue disease and autoimmune disease (e.g. RA, SLE, PAN, polymyalgia rheumatica); sarcoidosis.

Thrombosis DVT; PE.

Drugs e.g. antibiotics.

Fever in children <5y ➔ p. 852.

Sepsis ➔ p. 622

Rigors Shaking episodes (sometimes violent) associated with sudden rise in fever.

Night sweats Consider TB, lymphoma, leukaemia, solid tumour (e.g. renal carcinoma), menopause, anxiety states, drug causes, e.g. opioids, SSRIs.

Pyrexia of unknown origin Defined as a fever (either intermittent or continuous) which has lasted for >3wk and for which no cause has been found. Re-check history. Re-examine carefully. Check FBC; EBV screen (depending on age of the patient); ESR; CRP; LFTs; amylase; urine (M,C&S); viral titres (including HIV); blood cultures; and CXR. If cause does not become obvious refer urgently for further investigation.

Notifiable diseases (ND) Notification of certain diseases is required under the Public Health (Control of Disease) Act 1984 and Health Protection (Notification) Regulations 2010. Notification is made to the 'proper officer of the local authority' on forms available online (🖱 https://www.gov.uk/government/collections/notifications-of-infectious-diseases-noids). Diseases included:

- Acute encephalitis
- Acute infectious hepatitis
- Acute meningitis
- Acute poliomyelitis
- Anthrax
- Botulism
- Brucellosis
- Cholera
- COVID-19
- Diphtheria
- Enteric fever (typhoid/paratyphoid)
- Food poisoning
- Haemolytic uraemic syndrome
- Infectious bloody diarrhoea
- Invasive group A streptococcal disease and scarlet fever
- Legionnaire's disease
- Leprosy
- Malaria
- Measles
- Meningococcal septicaemia
- Mumps
- Plague
- Rabies
- Rubella
- SARS
- Smallpox
- Tetanus
- Tuberculosis
- Typhus
- Viral haemorrhagic fever
- Whooping cough
- Yellow fever

❗ In addition, notify other infections or contamination which could present significant risk to human health.

Sepsis

Sepsis is defined as life-threatening organ dysfunction caused by a dysregulated host response to an infection. It can be triggered by any infection. ~147,000 people/y are admitted to hospital with sepsis in the UK; 30% die (44,000 deaths/y). Prompt recognition and management saves lives.

Patients at high risk of sepsis

- Age <1y or >75y
- Frailty
- IV drug misuse
- Indwelling catheter/line
- Neonate—particularly if premature (<37wk), PROM, or maternal history of infection
- Immunosuppression, e.g. chemotherapy, steroids, or immunosuppressant drugs, asplenic/hyposplenic, sickle cell disease, DM
- Invasive procedure within 6wk, e.g. surgery, childbirth, miscarriage/TOP
- Breach of skin integrity, e.g. cut, ulcer, blister, burn

Initial assessment If a patient presents with possible infection, think '*Could this be sepsis?*'—particularly if the person looks ill or the family/carer is concerned.

History Ask about risk factors, Symptoms may give clues to source of infection (e.g. dysuria, coughing, cellulitis) or trigger concern (e.g. breathlessness, dizziness, cold peripheries, abnormal behaviour). Ask about frequency of urination in the past 18h. If assessing remotely, arrange to see if any risk factors or concerning features. Assess with extra care if communication problems, e.g. non-English speaker, dementia, dysphasia.

Examination Look for source of infection. Examine for mottled/ashen appearance; cyanosis; non-blanching rash; any breach of skin integrity (e.g. cut, burn, or skin infection) or other rash indicating potential infection. Check standard observations:

- Temperature
- BP (child—only if child cuff available)
- Heart rate
- Respiratory rate
- O2 saturation
- Conscious level
- Capillary refill time (children <5y)

Risk stratification

- Adults and children ≥12y—Figure 17.1
- Children aged 5–12y—Figure 29.12, ➲ p. 1058
- Children <5y ➲ p. 852

Management

- If any red flag (high-risk) features (Figure 17.1), admit via blue light ambulance to hospital. Give O_2 to maintain peripheral saturations >94% while awaiting transfer. Provide clear handover to paramedics (e.g. SBAR—➲ p. 1035; NEWS2 score—➲ p. 1037)
- If any amber flag features (Figure 17.1), assess whether can be safely managed in the community, e.g. consider admission if lives alone. If managed in the community, treat underlying infection as appropriate. Provide clear safety netting advice about when/how to seek further advice if there is any deterioration or concern. Consider arranging early follow-up. ❗ If <17y and any immune suppression, admit

Meningitis/meningococcal sepsis ➲ p. 1056

Neutropenic sepsis ➲ p. 626

Category	High-risk criteria	Moderate- to high-risk criteria	Low-risk criteria
History	Objective evidence of new altered mental state	History from patient, friend, or relative of new onset of altered behaviour or mental state History of acute deterioration of functional ability Impaired immune system (illness or drugs including oral steroids) Trauma, surgery, or invasive procedures in the last 6wk	Normal behaviour
Respiratory	↑ respiratory rate: ≥25 breaths/min New need for O_2 to maintain saturation >92% (or >88% if COPD)	↑ respiratory rate: 21–24 breaths/min	No high-risk or moderate- to high-risk criteria met
BP	Systolic BP: ≤90mmHg or >40mmHg below normal	Systolic BP: 91–100mmHg	No high-risk or moderate- to high-risk criteria met
Circulation/ hydration	↑ heart rate: >130 bpm Not passed urine in ≥18h (if catheterized, passed <0.5 mL/kg/h of urine)	↑ heart rate: 91–130bpm (pregnant ♀: 100–130 bpm) or new-onset arrhythmia Not passed urine in 12–18h (if catheterized, passed 0.5–1mL/kg/h of urine)	No high-risk or moderate- to high-risk criteria met
Temperature		Tympanic temperature <36°C	
Skin	Mottled or ashen appearance Cyanosis of skin, lips, or tongue Non-blanching skin rash	Signs of potential infection, including redness, swelling, or discharge at surgical site or breakdown of wound	No non-blanching rash

Figure 17.1 Risk stratification tool for adults and children ≥12y with suspected sepsis

Reproduced with permission from NICE (2016, updated 2017) Sepsis: recognition, diagnosis and early management. www.nice.org.uk/guidance/ng51

Further information

NICE (2016, updated 2017) Sepsis: recognition, diagnosis and early management. 🖱 www.nice.org.uk/guidance/ng51

Illness in returning travellers

⚠ In all unwell returning travellers, consider imported disease *in addition* to the usual differential diagnosis. Tropical medicine is a specialized field. If unsure seek expert advice by telephone or admit the patient.

History Ask about:

- Symptoms
- Areas travelled to (including brief stopovers)
- Duration of travel
- Immunizations prior to travel
- Malaria prophylaxis
- Health of members of the travel party
- Sexual contacts while abroad
- Medical treatment abroad

Examination Full examination. Particularly check for fever, jaundice, abdominal tenderness, chest signs, rashes, lymphadenopathy.

Investigations Depend on symptoms and examination findings. *Consider*: FBC, thick and thin blood films for malaria, LFTs, viral serology, blood culture, stool culture (ensure it is fresh), MSU.

Malaria 2000 cases/y are notified in the UK. Easy to miss.

- Symptoms Malaria is a great mimic and can present with virtually any symptoms. Usually consists of a prodrome of headache, malaise, myalgia, and anorexia followed by recurring high fevers, rigors, and drenching sweats—lasting 8–12h at a time
- Examination May be normal—look for anaemia, jaundice, ± hepatosplenomegaly
- Investigation In all cases of fever in patients who have returned from a malarial endemic area—even if the plane just landed there and they did not get off, send a thick and thin film for malaria
- Management Admit for further investigation and treatment if:
- Very unwell
- Unable to check a thick and thin film (e.g. presentation at a weekend or out of hours)
- Thick and thin film +ve
- Persistent fever despite −ve thick and thin film

Falciparum malaria Caused by *Plasmodium falciparum*. Accounts for ~½ UK cases—it may not present for up to 3mo after return from a malarial area. Can be fatal in <24h—especially if it occurs in pregnant women or small children (<3y). *Complications*: cerebral malaria (80% deaths); hypoglycaemia; renal failure; pulmonary oedema; splenic rupture; disseminated intravascular coagulation; death.

Benign malaria Caused by *P. vivax*, *P. ovale*, and *P. malariae*. May cause illness up to 18mo after return. All have very low mortality. Relapse may occur at intervals after initial infection as parasites lie dormant in the liver (*P. vivax* and *P. ovale*) or blood (*P. malariae*).

TyphoidND and paratyphoidND Caused by *Salmonella typhi* and *Salmonella paratyphi*, ~200 cases/y are notified in the UK.

- Spread By the faeco-oral route
- Incubation 3d—3wk

- Symptoms Usually malaise, fever, headache, cough, constipation (or diarrhoea), nose bleeds, bruising, and/or abdominal pain
- Examination Pyrexia; relative bradycardia; rose-coloured spots on the trunk (40%); splenomegaly; CNS signs (coma, delirium, meningism)
- Management Admit for further investigation and antibiotics
- Prognosis 10% die if untreated—<0.1% if treated; 1% become chronic carriers after infection

Dengue fever Viral infection endemic in tropical and subtropical regions. ~150 cases/y are notified in the UK.

- Spread By the day-biting *Aedes* mosquito
- Incubation 4–7d
- Symptoms/examination Usually presents with a flu-like illness, sudden high fever ± red, maculopapular rash (appears 2–5d after the fever). Other symptoms—fatigue, headache, arthralgia, myalgia, nausea and vomiting, lymphadenopathy, skin hypersensitivity
- Dengue haemorrhagic fever Rare, severe form of dengue fever with poor prognosis. Purpuric rash appears 2–3d after onset of symptoms; minor injuries may cause bleeding; shock → death. *Risk factors*: previous dengue infection, age <12y, ♀, Caucasian
- Management Admit; treatment is supportive

Traveller's diarrhoea 50% of travellers experience some diarrhoea. Most cases last 4–5d; 1–2% last >1mo. In all cases send a fresh stool sample for M,C&S at first presentation, noting on the form areas visited. Consider all the usual causes for diarrhoea (➔ p. 346) and gastroenteritis (➔ p. 348). In addition consider:

CholeraND Caused by Gram −ve bacterium *Vibrio cholerae*.

- Spread By faeco-oral route
- Incubation Few hours–5d
- Presentation Profuse watery stool, fever, vomiting, dehydration
- Management Admit. Treatment is with rehydration ± antibiotics

Giardiasis ~3800 cases/y are notified in the UK. Flagellate protozoan. Infection is suggested by an incubation period ≥2wk; watery stool with flatus ++ (explosive diarrhoea); no fever. Stool microscopy may be −ve. If suspected treat with metronidazole. Rapid response is diagnostic.

Amoebic dysenteryND ~100 cases/y are notified in the UK. May begin years after infection. Diarrhoea begins slowly becoming profuse and bloody ± fever ± malaise. Diagnosis is confirmed by microscopy of fresh stool. Take specialist advice on management.

Sexually transmitted infections ➔ p. 714

HIV ➔ p. 720

Hepatitis A ➔ p. 394

Hepatitis B and C ➔ p. 718

TB ➔ p. 296

Meningitis ➔ p. 1056

Infection and immunocompromise

Infections in patients whose host defence mechanisms are compromised range from minor to fatal. They are often caused by organisms that normally reside on body surfaces.

⚠ **Neutropenic sepsis** Defined as fever of ≥38°C for ≥2h when the neutrophil count is $<1.0 \times 10^9$/L. *Causes*:

- Chemotherapy (most common cause)
- Radiotherapy—if large volumes of bone marrow are irradiated
- Malignant infiltration of the bone marrow, e.g. prostate/breast cancer

Risk of neutropenia Risk of bacterial/fungal infection ↑ sharply as neutrophil count falls to $<1.0 \times 10^9$/L, with greatest risk at counts $<0.1 \times 10^9$/L. Neutropenia for >5d is a further risk factor. Patients are at most risk in the nadir period from 1wk after therapy.

Presentation Mouth ulcers and ↑ fatigue can be signs of neutropenia. Symptoms/signs of sepsis may be minimal and deterioration can be rapid. Development of fever in a patient with neutropenia is a medical emergency caused by infection until proven otherwise. Early referral for investigation and specialist management can be life-saving.

Opportunistic infections Infections from endogenous microflora that are non-pathogenic or from ordinarily harmless organisms. Occur if host defence mechanisms have been altered by:

- Age
- Infection
- Burns
- Neoplasms
- Metabolic disorders
- Irradiation
- Foreign bodies
- Corticosteroids
- Immunosuppressive or cytotoxic drugs
- Diagnostic or therapeutic instrumentation

The precise character of the host's altered defences determines which organisms are likely to be involved. These organisms are often resistant to multiple antibiotics. Organisms commonly involved:

- *E. coli*
- CMV
- *Pneumocystis*
- *Candida*
- Herpes viruses
- Toxoplasmosis
- Mycobacteria
- Non-pathogenic streptococci
- Cryptococcal infection

Expert care is always required—refer promptly to the consultant responsible for the patient.

HIV ➔ pp. 720–3

Asplenic patients All asplenic patients (or functionally asplenic patients, e.g. patients with sickle cell disease) are at ↑ risk of bacterial infection. Warn patients about severe malaria and other tropical infections when travelling abroad. Admit to hospital if infection develops despite prophylactic measures. Ensure that asplenic patients have:

- Vaccinations Table 17.3
- Prophylactic antibiotics Oral penicillin continuously until age 16y or for ≥2y post-splenectomy—whichever is longer
- Stand-by amoxicillin To start if symptoms of infection begin
- Patient held card Alerting health professionals to infection risk—cards and information leaflets for patients are available from 🖱 www.orderline.dh.gov.uk ☎ 0300 123 1002

Table 17.3 Immunization schedule for children and adults with immunocompromise, complement deficiency, asplenia, or splenic dysfunction

Age at presentation	Vaccination schedule
<2y	• Routine childhood vaccination schedule (➔ p. 619) • Further booster dose of MenACWY >1mo after the 12–13mo routine Hib/MenC and PCV13 vaccination • 1 further dose of Hib/MenC and PPV at >2y of age
2–5y	• If already vaccinated with PCV7, vaccinate with PCV13 and then, after ≥2mo, with PPV • If already vaccinated with PCV13, vaccinate with PPV • Vaccinate immediately with Hib/MenC booster • 1mo after Hib/MenC booster, vaccinate with MenACWY
>5y	• Give Hib/MenC and PPV immediately • 1mo after Hib/MenC and PPV, vaccinate with MenACWY

PCV7—pneumococcal conjugate 7-valent vaccine
PCV13—pneumococcal conjugate 13-valent vaccine
PPV—pneumococcal polysaccharide vaccine
Hib/MenC—*Haemophilus influenza* b/meningitis conjugate vaccine
MenACWY—meningitis quadrivalent (ACWY) conjugate vaccine

Prevention of infection

Antibiotics Used for prevention of:

- TB and meningitis in exposed patients
- Streptococcal infection in asplenic/hyposplenic patients
- Bacterial infections in granulocytopenic patients
- Pneumocystis in AIDS patients
- Recurrent UTIs

⚠ Watch for signs of superinfection with resistant organisms.

Active immunization Table 17.3

Passive immunization Can prevent or ameliorate herpes zoster (VZ-Ig), hepatitis A and B, measles, and cytomegalovirus infection in selected immunosuppressed patients. If a patient is in contact with any of these diseases, ask for advice from the consultant looking after the patient or a consultant in communicable disease control.

Immunoglobulin administration Effective for patients with hypogammaglobulinaemia. Given on a regular basis by IV infusion.

❗ Start vaccinations ≥2wk before splenectomy/starting immunosuppressive treatment.

Further information

Public Health England (2014) The Green Book: Immunisation against infectious disease. 🖱 www.gov.uk/government/collections/immunisation-against-infectious-disease-the-green-book

Childhood viral infections

Management For infections listed in Table 17.4, management is supportive with paracetamol, fluids ± antibiotics for 2° infection. Teething gels, e.g. Calgel®, may sooth mouth lesions in hand, foot, and mouth disease. Admit if any serious complications develop.

Prevention of measles, mumps, and rubella Measles, mumps, and rubella (MMR) vaccination consists of live attenuated measles, mumps, and rubella viruses. Vaccine viruses are not transmitted.

- Offer MMR to all children after their 1st birthday and again preschool. Re-immunization is needed if given to children of <1y. Children with chronic illness, e.g. CF, are at particular risk from measles and should be immunized. Malaise, fever, and rash are common ~1wk after immunizations and last 2–3d. Advise on fever management. There is no link between MMR and autism or inflammatory bowel disease
- Offer children aged >18mo who have not been vaccinated (or whose vaccination status is unclear) 2 doses of MMR vaccine ≥1mo apart; if the child has received 1 dose of MMR, give a booster dose
- Offer MMR to women of child-bearing age who are not immune to rubella (i.e. have not had 2 doses of MMR or are seronegative). ❗ There is no evidence that vaccination in pregnancy is harmful, but do not give to women known to be pregnant and advise women who are vaccinated to avoid pregnancy for 1mo afterwards

Prevention of chickenpox Offer chickenpox (varicella) immunization (2 doses 4–8wk apart) to non-immune healthcare workers who have direct patient contact, and susceptible close contacts of immunocompromised patients, where continuing contact is unavoidable. Consider those with a definite history of varicella infection immune—antibody test others. Vaccination is contraindicated if pregnant or immunocompromised.

Non-immune, immunosuppressed patients (➔ p. 626), pregnant women (➔ p. 794), or neonates (➔ p. 795) with significant exposure to chickenpox/shingles, should receive zoster immunoglobulin (VZ-Ig) <3d after contact. Check antibody levels if immune status is unknown.

Shingles Re-inactivation of latent chickenpox virus; shingles cannot be acquired by exposure to chickenpox but contacts of patients with shingles can develop chickenpox. Infectious until all lesions have scabbed.

- **Incidence** 1 in 25. Any age—more common if immunocompromised
- **Presentation** Unilateral pain precedes a vesicular rash by 2–3d. Crops of vesicles appear over 3–5d and are in the distribution of ≥1 adjacent dermatomes. The affected area is usually hyperaesthetic—pain may be severe. Lesions scab over and fall off in <14d
- **Management** Treat as for chickenpox. Oral antivirals (e.g. aciclovir 800mg 5×/d) are only effective if initiated <48h after onset of the rash. If immunocompromised admit for IV antivirals
- **Prevention** Shingles vaccination is routinely offered to adults aged >70y in the UK

Complications Postherpetic neuralgia (➔ p. 188); dissemination to other areas (immunosuppressed patients—admit for IV antivirals); eye involvement—refer urgently to ophthalmology; Ramsay Hunt syndrome (➔ p. 512).

Table 17.4 Common childhood viral infections

Condition	Duration	Main symptoms
MeaslesND	10d	*Incubation*: 10–14d *Early symptoms*: fever, conjunctivitis, cough, coryza, LNs *Later symptoms*: Koplik's spots (tiny white spots on bright red background found on buccal mucosa of cheeks), rash (florid maculopapular appears after 4d—becomes confluent) *Complications*: bronchopneumonia, otitis media, stomatitis, corneal ulcers, gastroenteritis, appendicitis, encephalitis (1 in 1000 affected children), subacute sclerosing panencephalitis (rare)
RubellaND *(German measles)*	10d	*Incubation*: 14–21d *Symptoms*: mild and may pass unrecognized. Fever, LNs (including suboccipital nodes), pink maculopapular rash which lasts 3d *Complications*: birth defects if infected in pregnancy; arthritis (adolescents); thrombocytopenia (rare); encephalitis (rare)
MumpsND	10d	*Incubation*: 16–21d *Symptoms*: subclinical infection is common. Fever, malaise, tender enlargement of 1 or both parotids ± submandibular glands *Complications*: aseptic meningitis; epididymo-orchitis; pancreatitis
Chickenpox	14d	*Incubation*: 10–21d (infectious 1–2d before rash appears and for 5d afterwards) *Symptoms*: rash ± fever. Spots appear in crops for 5–7d on skin and mucus membranes and progress from macule → papule → vesicle then dry and scab over *Complications*: eczema herpeticum (➔ p. 578); encephalitis (cerebellar symptoms most common); pneumonia; birth defects; neonatal infection (➔ p. 795)
Roseola infantum	4–7d	Child <2y *Symptoms*: high fever, sore throat, and lymphadenopathy, macular rash appears after 3–4d when fever ↓
Erythema infectiosum (5th disease/ slapped cheek)	4–7d	Parvovirus infection *Symptoms*: erythematous maculopapular rash starting on the face ('slapped cheeks'), reticular, 'lacy' rash on trunk and limbs, mild fever, arthralgia (rare) Contact with parvovirus in pregnancy—➔ p. 792
Hand, foot, and mouth disease	5–7d	Coxsackie virus infection *Symptoms*: oral blisters/ulcers, red-edged vesicles on hands and feet, mild fever

Streptococcal and staphylococcal infections

Streptococcal infection Several groups are pathogenic to man—A, B, C, G, D, and viridans streptococci. Presentation is varied:

- Pharyngitis
- Tonsillitis
- Wound/skin infections
- Septicaemia
- Scarlet fever
- Pneumonia
- Rheumatic fever
- Glomerulonephritis
- Neonatal sepsis
- Postpartum sepsis
- Endocarditis
- Septic arthritis
- Pneumonia
- UTI
- Dental caries

Investigation Diagnosis is usually clinical. Evidence of infection can be gained by measuring changing antibody response (ASO titres). ASO titres are ↑ in ~80% infections. Wound swabs are +ve if infection is on the skin and throat swabs may be +ve in pharyngitis/tonsillitis.

Treatment Most streptococci are sensitive to penicillin (e.g. phenoxymethylpenicillin 250–500mg qds for 7–10d) although resistance is increasingly common.

Pneumococcal infection There are >85 types of *S. pneumoniae*. Pneumococci are carried in the noses and throats of ½ the population. In most people they are harmless. Spread is by droplet infection.

Presentations

- Pneumonia
- Acute otitis media
- Sinusitis
- Meningitis
- Endocarditis
- Septic arthritis (rare)
- Peritonitis (rare)

Treatment Amoxicillin 250–500mg tds for 7d (clarithromycin in allergic individuals). Resistance to penicillin in the community is still low.

Childhood vaccination Routine vaccination is offered as part of childhood immunization programme (➔ p. 619) using 13-valent pneumococcal conjugate vaccine (PCV) given at 2mo, 4mo, and 12–13mo of age.

Vaccination of high-risk groups A single dose of pneumococcal polysaccharide vaccine (PPV) is indicated for high-risk patients (Box 17.1) who have not previously been vaccinated. Special rules apply to patients who are immunosuppressed or have deficient spleens (Table 17.3 ➔ p. 627). Booster doses are not needed except for patients with asplenia or nephrotic syndrome—when give a booster after 5–10y.

Scarlet fever Group A haemolytic streptococcal infection. *Incubation*: 2–4d. Presents with fever, malaise, headache, tonsillitis, rash (fine punctate erythema sparing face, 'scarlet' facial flushing), and strawberry tongue (initially white turning red by day 3–4). Treat with phenoxymethylpenicillin 250–500mg qds for 10d (or clarithromycin if allergic). Complications include rheumatic fever (➔ p. 246) and acute glomerulonephritis (➔ p. 416).

Staphylococcal infection Usually *Staph. aureus*—occasionally *Staph. epidermidis*. Carried in the nose of ~30% of healthy adults. Antibiotic-resistant strains are common

Box 17.1 High-risk patients for pneumococcal infection

Those:

- ≥65y of age
- With coeliac disease
- With a cochlear implant
- With asplenia/functional asplenia (e.g. splenectomy, sickle cell)
- With immune deficiency due to disease (e.g. lymphoma, Hodgkin's disease, multiple myeloma, HIV) or treatment (e.g. chemotherapy, prolonged systemic steroids)
- With chronic heart disease, lung disease (e.g. asthma, COPD), renal disease (or nephritic syndrome), or liver disease
- With DM requiring insulin or oral hypoglycaemic drugs, and/or
- With CSF shunt/other conditions where leakage of CSF fluid can occur

Presentation

- Breast abscess/mastitis
- Abscesses/furuncles/carbuncles
- Septicaemia
- Endocarditis
- Wound infection
- Osteomyelitis/septic arthritis
- Pneumonia—especially patients with COPD, influenza, or those receiving corticosteroids or immunosuppressive therapy
- Neonatal infections—usually appear <6wk after birth—pustular or bullous skin lesions on neck, axilla, or groin

Management Antibiotics (usually flucloxacillin 250–500mg qds or clarithromycin 250–500mg bd for 7–10d), abscess drainage where appropriate, and general supportive measures. Where possible, obtain specimens for culture before instituting or altering antibiotic regimens.

Staphylococcal scalded skin syndrome ➔ p. 883

Meticillin-resistant *Staph. aureus* (MRSA) MRSA acts in the same way as any other *Staph. aureus*—it is carried harmlessly in most but occasionally causes a range of infections. It is only different because of its multiple resistance to antibiotics. Often contracted in hospital.

- ↓ tendency for multiple resistance by prudent use of antibiotics
- Wash hands thoroughly with an appropriate antibacterial preparation if they appear soiled
- If hands appear clean, wash with an alcoholic rub between each and every patient contact
- Follow local policies for management of patients who are known to be infected with or carry MRSA

Toxic shock syndrome Caused by staphylococcal exotoxin.

- Risk factors Tampon use; postpartum; staphylococcal wound infection; influenza; osteomyelitis; cellulitis
- Presentation Sudden-onset high fever, vomiting, diarrhoea, confusion, and skin rash. May progress to shock ± death
- Management Admit as a medical emergency—mortality 8–15%

Further information

National Electronic Library for Infection Antimicrobial resistance website.
🖱 www.antibioticresistance.org.uk

Other bacterial infections

Haemophilus influenzae

Haemophilus influenza type b (Hib) Vaccination against Hib is routinely offered to all children (➔ p. 619) and immunocompromised patients (➔ p. 627). Prior to routine vaccination, Hib infection accounted for 60% of meningitis in children aged <5y. It was also a common cause of epiglottitis, septicaemia, and septic arthritis/osteomyelitis. Infection is now rare.

Other types Non-encapsulated *Haemophilus influenzae* (ncHi) usually causes non-invasive respiratory tract infections, e.g. otitis media, sinusitis. It can cause invasive disease in neonates (<1mo of age) and those with co-morbidities, e.g. malignancy, immunosuppression, DM, chronic renal/liver/lung disease. Other *H. influenzae* serotypes (Hia, Hic, Hid, Hie, Hif) only rarely cause invasive disease.

Management Organisms are often penicillin resistant. If severe infection, admit.

Clostridium infections Anaerobic, spore-forming bacilli found in dust, soil, vegetation, and GI tracts of humans and animals. 25–30 species cause disease in humans. *Presentations*:

- Food poisoning—*C. perfringens*
- Pseudo-membranous colitis—overgrowth of *C. difficile* following antibiotic therapy—presents with bloody diarrhoea. Treated with vancomycin or metronidazole if toxin is isolated from stool
- Botulism—caused by a toxin released by *C. botulinum* which is ingested in contaminated food—presents with neurological symptoms and warrants immediate admission for antitoxin
- Wound infections—*C. perfringens* causes cellulitis which may → gas gangrene, septicaemia ± death—admit for IV antibiotics
- Tetanus

Meningitis ➔ p.1056

Tetanus (lockjaw)$^{\text{ND}}$ There are ~6 cases of tetanus and 1 death every year in the UK. *Incubation*: 2–50d. *C. tetani* infects contaminated wounds that may be trivial, the uterus postpartum (maternal tetanus), or newborn umbilicus (tetanus neonatorum). Tetanus prone injuries—Box 17.2.

Tetanus toxin causes generalized or localized tonic spasticity ± tonic convulsions. Suspect in any patient not immunized who develops muscle stiffness or spasm several days after suffering a skin wound or burn.

Prevention Tetanus vaccine:

- Primary immunization 3 doses with 1mo between each (i.e. 2nd dose 1mo after 1st dose and 3rd dose 1mo after 2nd dose)
- Booster doses 1 booster dose 5y after primary immunization (3y if child aged <10y) and a 2nd booster dose 10y after the 1st booster dose
- Open wounds ➔ p. 1089

Box 17.2 Tetanus-prone injuries

- Puncture-type injuries acquired in a contaminated environment and likely to contain tetanus spores, e.g. gardening injuries
- Wounds containing foreign bodies, e.g. splinters
- Compound fractures
- Wounds/burns with systemic sepsis
- Certain animal bites and scratches from animals that have been rooting in soil, or agricultural animals

High tetanus risk Heavy contamination with material likely to contain tetanus spores; extensive devitalized tissue and/or wounds awaiting surgical intervention delayed for >6h.

DiphtheriaND Caused by *Corynebacterium diphtheriae*. Rare in the UK since routine immunization. Spread by droplet infection, contact with articles soiled by an infected person. *Incubation*: 2–5d.

Presentation In countries where hygiene is poor, cutaneous diphtheria is the predominant form. Elsewhere, characterized by an inflammatory exudate which forms a greyish membrane in the respiratory tract (may cause respiratory obstruction). *C. diphtheriae* secretes a toxin which affects myocardium, nervous, and adrenal tissues.

Management Admit for antitoxin and IV antibiotics. Patients may be infectious for up to 4wk but carriers shed *C. diphtheriae* for longer.

Prevention Vaccination—given routinely in childhood in the UK (➔ p. 619). In addition give booster dose to people in contact with a patient with diphtheria or carrier, or before travel to epidemic or endemic areas.

Pseudomonas aeruginosa Common. Treatment is difficult due to multiple antibiotic resistance—if suspected send specimen for M,C&S.

- In immunocompetent patients may cause UTI, wound infections (particularly leg ulcers—gives a characteristic greenish colouring), osteomyelitis, and skin infections (e.g. otitis externa)
- In immunocompromised patients and patients with CF, it is a common cause of pneumonia and septicaemia

Enterobacteria Examples include:

- *Salmonella*
- *Shigella*
- *Escherichia*
- *Klebsiella*
- *Enterobacter*
- *Proteus*
- *Morganella*
- *Providencia*
- *Yersinia*

Some are normal gut commensals. Others are pathogenic causing:

- Diarrhoea and intra-abdominal infections—e.g. peritonitis, hepatobiliary
- UTI—often *E. coli; Proteus* species are associated with bladder stones
- Septicaemia and/or meningitis—*E. coli* is the most common cause of meningitis in neonates
- Chest infection—*Klebsiella* may cause a severe form of pneumonia
- Endocarditis—rare

Organisms are usually sensitive to trimethoprim. Severe infection requires admission to hospital for IV antibiotics.

Chapter 18

Haematology and immunology

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Full blood count and ESR

The most commonly requested blood test is the full blood count (FBC).

❗ Reference ranges are a guide only; normal values are affected by ethnicity, age, and pregnancy. For mild abnormalities in well individuals, take laboratory advice to avoid over-investigation/referral.

Red cells Anaemia→ p. 638; erythrocytosis→ p. 655

Mean cell volume (MCV)

- ↓ MCV (microcytic; <80fL): iron-deficiency anaemia. Confirm with ↓ serum ferritin. *Rarer causes*: thalassaemia (suspect if MCV is 'too low' for level of anaemia); congenital sideroblastic anaemia (very rare)
- ↑ MCV (macrocytic; >100fL): vitamin B_{12}/folate deficiency; alcohol; liver disease; drugs (e.g. azathioprine, chemotherapy); haemolysis; pregnancy; hypothyroidism; marrow infiltration; myelodysplasia

White cells Normal white cell count is 4–11 × 10^9/L. Reasons for individual components to be ↑ or ↓ are summarized in Table 18.1.

Platelets

Thrombocytopenia ↓ platelets (<150 × 10^9/L). *Causes*:

- ↓ production Marrow failure; alcohol; megaloblastic anaemia
- ↓ survival Pregnancy; ITP; viruses; disseminated intravascular coagulation; SLE; lymphoma; thrombotic thrombocytopenic purpura; hypersplenism; genetic disease
- Platelet aggregation Heparin (5% patients)
- Drugs e.g. omeprazole, furosemide, quinine, trimethoprim

Thrombocytosis ↑ platelets (>450 × 10^9/L). *Causes*:

- 1° thrombocytosis
- Reactive (2°) thrombocytosis Due to infection, malignant disease, acute or chronic inflammatory disease, pregnancy, after splenectomy, iron deficiency, or following haemorrhage

> ❗ ~50% with persistent, unexplained thrombocytosis have a malignancy. Consider[N]:
>
> - Urgent CXR if ≥40y
> - Urgent pelvic USS if ♀, age ≥55y, + vaginal discharge/haematuria
> - Routine referral for upper GI endoscopy if aged ≥55y + nausea, vomiting, weight ↓, reflux, dyspepsia, and/or upper abdominal pain

Pancytopenia ↓ in red cells, white cells, and platelets. *Causes*:

- Aplastic or megaloblastic anaemia
- Bone marrow infiltration or replacement, e.g. by lymphoma, leukaemia, myeloma, 2° carcinoma, myelofibrosis
- Hypersplenism
- Disseminated TB
- SLE
- Paroxysmal nocturnal haemoglobinuria

Erythrocyte sedimentation rate (ESR) Rate of fall of red cells in a column of blood. A measure of the acute phase response—the pathological process may be infective, immunological, malignant, ischaemic, or traumatic. Normal values ↑ with age; ♀ > ♂. ❗ ↑ in patients with severe anaemia.

Table 18.1 Differential diagnosis for white cell count changes

White cell type *Normal range* (%)	Causes of ↑ count	Causes of ↓ count
Neutrophils *$2.0–7.5 \times 10^9/L$* (40–75%)	- Bacterial infection - Physical injury, e.g. trauma, burns, surgery - Inflammation, e.g. PMR, RA - Myocardial infarction - Pregnancy - Malignancy—leukaemia, disseminated malignancy - Drugs, e.g. steroids	*Mild $1–2 \times 10^9/L$* - Viral infections, e.g. mumps, hepatitis, influenza - Drugs, e.g. carbimazole, cytotoxics - Idiopathic/immune - Benign ethnic (Afro-Caribbean) *If $<1 \times 10^9/L$* discuss with haematology + refer. Warn about risks of neutropenic sepsis (➔ p. 626) *If $<0.5 \times 10^9/L$* refer urgently to haematology
Lymphocytes *$1.5–4.9 \times 10^9/L$* (20–45%)	- Viral infection, e.g. EBV, early HIV, hepatitis, rubella - Other infections—whooping cough, toxoplasmosis - CLL and ALL ❗ Large numbers of abnormal (‘atypical’) lymphocytes are characteristically seen with EBV infection.	- Drugs, e.g. cytotoxics, steroids - Systemic disease, e.g. influenza, SLE, uraemia - HIV infection ❗ Unless other haematological abnormalities, never needs haematology referral
Monocytes *$0.2–0.8 \times 10^9/L$* (2–10%)	- Chronic bacterial infections (e.g. TB, SBE) - Autoimmune disorders	N/A
Eosinophils *$0.04–0.44 \times 10^9/L$* (1–5%) ❗ If mildly ↑ ($<1.5 \times 10^9/L$) in well patient—do not investigate further	- Atopic disease, e.g. asthma (80%) - Parasitic infections (8%) - Haematological malignancy (2.5%) - Allergic/atopic skin conditions (2%) - Solid tumours (2%) - GI disease (inflammatory bowel disease; coeliac) 1.5% - Lung disease (1%) - Connective tissue disease (0.5%)	N/A
Basophils *$0.01–0.1 \times 10^9/L$* (<1%)	- CML/myeloproliferative disease - Hypothyroidism - Drugs, e.g. oestrogen	N/A

Further information

NICE (2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Anaemia: diagnosis and initial investigation

Anaemia Anaemia is a lack of sufficient red blood cells and thus haemoglobin (♂: Hb <13g/dL; ♀: Hb <12g/dL or <11g/dL 1st trimester and <10.5g/dL 2nd/3rd trimester of pregnancy). It results if there is:

- ↓ red cell production Defective precursor proliferation/maturation
- ↑ loss or rate of destruction Bleeding or haemolysis
- ↓ tissue requirement for oxygen Usually hypothyroidism

Presentation Patients who become anaemic slowly may remain asymptomatic for a long time. As anaemia progresses, pallor, exertional dyspnoea, tachycardia, palpitations, angina (especially if past history of coronary artery disease), night cramps, and cardiac bruits appear. Ultimately, with severe anaemia, high-output cardiac failure may develop.

❗ *Pallor* may indicate anaemia but is a very non-specific sign which may also be racial, familial, or cosmetic. *Other causes of pallor*: shock, Stokes–Adams attack, vasovagal faint, myxoedema, hypopituitarism, albinism.

Initial investigation of anaemia Table 18.2

Management Treat the cause—if no cause is found, refer for further investigation.

Table 18.2 Investigation and differential diagnosis of anaemia

MCV	Causes	Potential further investigations
Low <80fL	Iron deficiency Haemoglobinopathy (thalassaemia) Anaemia of chronic disorder	Blood film Reticulocyte count Ferritin CRP/ESR/plasma viscosity Hb electrophoresis (if indicated)
Normal	Acute blood loss Haemolysis Anaemia of chronic disorder Uraemia Haemoglobinopathy Marrow failure	Blood film Reticulocyte count Hb electrophoresis (if indicated) Ferritin Serum vitamin B_{12} Serum and red cell folate Renal function Serum bilirubin
High >100fL	Folate and/or vitamin B_{12} deficiency Alcohol Liver disease Thyroid disease Myelodysplasia/marrow infiltration	Blood film Serum vitamin B_{12} Serum and red cell folate Liver function Thyroid function tests

Further information

British Society for Haematology Diagnosis of B_{12} and folate deficiency. www.b-s-h.org.uk/guidelines

Vitamin B_{12} deficiency Vitamin B_{12} is found in liver, kidney, fish, chicken, meats, and dairy products. Absorption takes place by active and passive mechanisms—the latter being dependent on intrinsic factor, a protein produced by gastric parietal cells. Deficiency presents with macrocytosis, or with symptoms/signs: glossitis; mouth ulcers; peripheral neuropathy; ataxia; optic atrophy; memory loss; subacute combined degeneration of the cord; rarely psychosis.

❗ B_{12} levels correlate poorly with deficiency, especially in pregnancy. If strong clinical suspicion but normal B_{12} levels, discuss with haematology.

Causes of deficiency

- Inadequate dietary intake e.g. vegans
- Malabsorption GI disease/surgery—gastric (e.g. *H. pylori* infection; pernicious anaemia; gastrectomy), pancreatic (e.g. chronic pancreatitis), or ileal (e.g. coeliac disease—➔ p. 382; ileal resection)
- Drugs Metformin; drugs causing achlorhydria, e.g. H_2 antagonists; PPIs, COC may ↓ B_{12} levels due to ↓ carrier protein not deficiency

Pernicious anaemia Autoimmune condition associated with gastric atrophy and intrinsic factor (50%)/gastric parietal cell (85%) antibodies. *Risk factors*: FH, other autoimmune disease (e.g. vitiligo, hypothyroidism), premature greying, blood groups A and HLA3. Associated with ↑ risk of stomach and colorectal cancer, particularly if <2y after diagnosis.

Management Treat with vitamin B_{12} (hydroxocobalamin IM—1mg 3×/wk for 2wk then every 2–3mo). Confirm response by repeating FBC after 2 and 8wk. Consider oral B_{12} supplements if borderline results and non-specific symptoms. Consider referral to exclude underlying GI cause.

Folate deficiency Folate is found in highest concentrations in liver and yeast but is also in spinach, other green vegetables, and nuts. Deficiency presents with macrocytosis; symptoms/signs of anaemia ± polyneuropathy or dementia, mouth ulcers. Check vitamin B_{12}—deficiencies may coexist.

Causes of deficiency

- Inadequate dietary intake Common—e.g. old age, poor social conditions, malignancy, anorexia, excess alcohol (particularly spirits)
- Malabsorption Coeliac disease, Crohn's disease, partial gastrectomy, tropical sprue, lymphoma, diabetic enteropathy
- Excess use Pregnancy, lactation, prematurity, ↑ cell turnover (e.g. malignancy, haemolysis)
- Drugs Anticonvulsants, trimethoprim

Management Treat the cause. Supplement with folic acid 5mg od for 4mo. If malabsorption, may need to ↑ dose to 15mg od. Treat vitamin B_{12} deficiency. For prophylaxis in chronic haemolytic states or for renal dialysis, up to 5mg od long term is used (take specialist advice).

⚠ **Folate supplements in pregnancy** Advise women to take supplements from planning pregnancy to 12wk gestation to prevent neural tube defect. *Dose*: to prevent first occurrence—400mcg od; if FH of neural tube defect, *or* mother has BMI >30kg/m^2, DM, sickle cell anaemia or coeliac disease, *or* if taking anticonvulsants, *or* to prevent recurrence—5mg od.

Iron deficiency anaemia

Most common form of anaemia in the UK (prevalence 2–5% among adult men and post-menopausal women). Red blood cells are microcytic (↓ MCV) and hypochromic. Low serum ferritin and/or low transferrin saturation confirms iron deficiency. Exclude haemoglobinopathy (➔ p. 644) in those with FH or from at-risk ethnic groups.

Causes ❗ >1 cause may be present.

- **GI blood loss** Aspirin/NSAID use (10–15%); colonic carcinoma (5–10%); gastric carcinoma (5%); benign gastric ulceration (5%); angiodysplasia (5%); oesophagitis (5%); oesophageal carcinoma (1–2%)
- **Malabsorption** Coeliac disease (4–6%)—may be the presenting feature; post-gastrectomy; *H. pylori* colonization; gut resection
- **Non-GI blood loss** Menstruation (20–30%); blood donation (5%); haematuria (1%); epistaxis
- **Other** Pregnancy; lactation; premature infants; deficient diet

Investigation and management See Figure 18.1. Treat underlying cause where possible. Give iron supplements, e.g. ferrous sulphate 200mg bd po—may turn stools black and cause constipation. If not tolerated, try lower dose or alternative preparation, e.g. ferrous fumarate. Fruit juice rich in vitamin C taken with the iron may ↑ iron absorption. Hb should ↑ by 1g/dL/wk—confirm response 2–3wk after starting treatment. Continue treatment for 3mo after correction of deficiency to replenish iron stores.

> ⚠ Asymptomatic colonic/gastric cancer may present with iron deficiency anaemia. Refer via suspected cancer pathway[N] if iron deficiency PLUS >60y or rectal bleeding and >50y.
>
> Use quantitative faecal immunochemical tests for those who do not meet these criteria. Refer via suspected colorectal cancer pathway where tests show occult blood in the faeces[N].

Failure to respond to iron supplements Consider *H. pylori* (test/treat), coeliac disease (oral iron not absorbed), continuing bleeding, non-compliance with iron, diagnosis is incorrect or anaemia mixed.

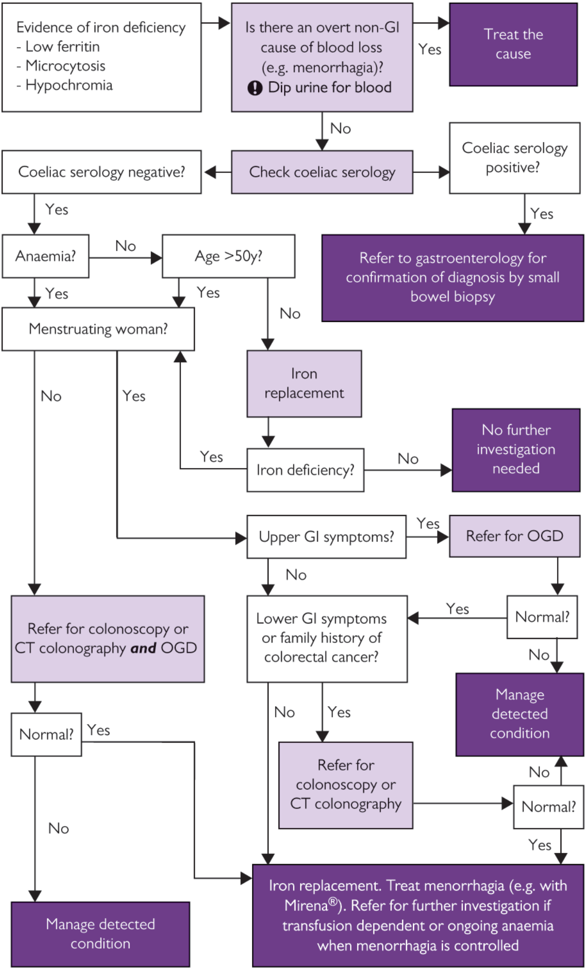
Follow-up Once blood count is normal, monitor Hb, MCH, and MCV every 3mo for 1y and then annually. Give further iron supplements if Hb, MCH, or MCV falls below normal levels. Investigate further if unable to maintain Hb.

Iron deficiency without anaemia Low serum ferritin (hypoferritinaemia) is 3× as common as iron deficiency anaemia but there is a very low prevalence of GI malignancy in this group (<1%). Investigate as in Figure 18.1. Give iron supplements to replenish iron stores.

Further information

British Society of Gastroenterology (2011) Guidelines for the management of iron deficiency anaemia. 🖱 https://www.bsg.org.uk/resource/guidelines-for-the-management-of-iron-deficiency-anaemia.html

NICE (2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12


Evidence of iron deficiency
- Low ferritin
- Microcytosis
- Hypochromia
Is there an overt non-GI cause of blood loss (e.g. menorrhagia)?
❗ Dip urine for blood
Yes
Treat the cause
No
Check coeliac serology
Coeliac serology negative?
Coeliac serology positive?
Yes
Yes
Refer to gastroenterology for confirmation of diagnosis by small bowel biopsy
Anaemia?
No
Age >50y?
Yes
Yes
No
Menstruating woman?
Iron replacement
No
Yes
Yes
Iron deficiency?
No
No further investigation needed
Upper GI symptoms?
Yes
Refer for OGD
No
Yes
Normal?
Refer for colonoscopy or CT colonography *and* OGD
Lower GI symptoms or family history of colorectal cancer?
No
Manage detected condition
Normal?
Yes
No
Yes
Refer for colonoscopy or CT colonography
No
Normal?
No
Yes
Iron replacement. Treat menorrhagia (e.g. with Mirena®). Refer for further investigation if transfusion dependent or ongoing anaemia when menorrhagia is controlled
Manage detected condition

❗ Referrals for colonoscopy/CT colonography should be 'urgent' i.e. to be seen in <2wk if age >60y, age>50y and rectal bleeding, or faecal blood test +ve[N]

Figure 18.1 Investigation and management of iron deficiency

Other anaemias

Anaemia of chronic disease (anaemia of inflammation)

Inflammatory cytokines suppress bone marrow erythropoiesis. Usually normocytic anaemia but can be microcytic if severe. Consider:

- Leg ulcers
- Cancer
- Old age
- Multi co-morbidities
- DM
- Low eGFR contributes via low erythropoietin

Management

- Aim to exclude significant disorders, e.g. haematological malignancy. Suitable tests include iron (↓), transferrin (↓), ferritin (normal or ↑), LDH, serum electrophoresis, eGFR, glucose, LFTs
- Optimize management of co-morbidities

❗ Ageing is pro-inflammatory process—50% of anaemia of the elderly is unexplained after investigation.

Myelodysplastic syndrome (MDS)

Common clonal, preleukaemia disorder affecting 1 in 500 >65y. Very variable severity from mild anaemia for many years to severe pancytopenia progressing to acute leukaemia within months. ❗ Isolated macrocytosis can precede anaemia for many years.

Causes Previous chemotherapy, chemical exposure, age.

Presentation Anaemia (usually macrocytic), neutropenia, thrombocytopenia, monocytosis, pancytopenia, infection, and/or bleeding. No splenomegaly or lymphadenopathy except in chronic myelomonocytic leukaemia (CMML).

Management Principally supportive—blood transfusion, erythropoietin, treat infections. Chemo- or immunosuppressive therapy is used in some situations. Stem cell transplant can be curative in young, fit patients; most patients are elderly.

Prognosis Depends on cytopenias, bone marrow cytogenetics, and bone marrow blast count.

Vitamin B_{12} and folate deficiency ➔ p. 639

Haemoglobinopathy ➔ p. 644

Haemolysis Normal red cells survive 120d before being removed from the circulation—mainly by the spleen. In haemolytic anaemia, red cells are destroyed faster than they are produced and anaemia develops.

Presentation May have FH. Symptoms/signs of anaemia may be accompanied by jaundice due to bilirubin released when red cells are destroyed; there may be splenomegaly ± haemoglobinuria. *FBC*: ↓ Hb; ↑ reticulocytes. Film shows polychromasia ± abnormally shaped cells (e.g. spherocytes) or other clues as to the cause of the haemolysis (e.g. fragmented cells suggest mechanical damage). Can be exacerbated by intercurrent illness, e.g. aplastic crisis with parvovirus.

Causes Table 18.3

Management Refer to haematology for advice. If possible, stop drugs that may be contributing. Treatment depends on cause, and may include steroids or immunosuppressants. All require folate supplements; thromboprophylaxis may be indicated for severe cases.

Aplastic anaemia^{G} Bone marrow failure characterized by pancytopenia. Rare, affecting 2–3/million population. No cause is found in 70–80%. Identified causes include genetic; drugs (inform Medicines and Healthcare products Regulatory Agency—➔ p. 120); toxins; infection.

Presentation Anaemia (➔ p. 638), thrombocytopenia (➔ p. 636), and infection resulting from neutropenia (➔ p. 626—less common). FBC reveals pancytopenia and lack of reticulocytes.

Management Refer urgently to haematology. Treatment is:

- Supportive Transfusions and antibiotics; or
- Definitive Restores a working bone marrow. Options depend on age and disease severity. Immunosuppression or stem cell transplant

Table 18.3 Causes of haemolytic anaemia

Cause	Examples
Congenital	
Membrane abnormalities	Hereditary spherocytosis or elliptocytosis
Haemoglobin abnormalities	Abnormal Hb, e.g. sickle cell anaemia—➔ p. 645 Defective synthesis, e.g. thalassaemia—➔ p. 644
Metabolic abnormalities	Glucose-6-phosphate dehydrogenase (G6PD) or pyruvate kinase deficiency
Acquired	
Immune	Autoimmune: • Warm, e.g. 2° to SLE, CLL, or NHL • Cold, e.g. 2° to EBV or mycoplasma infection Isoimmune (e.g. transfusion reaction; haemolytic disease of the newborn—➔ p. 770) Drug induced
Hypersplenism	Malaria, lymphoma, RA, portal hypertension
Red cell fragmentation	Artificial heart valves
Activated complement	Paroxysmal nocturnal haemoglobinuria
Secondary	Renal disease, liver disease
Miscellaneous	Infection (e.g. malaria), burns, chemicals, toxins, drugs

Further information

British Society for Haematology 🔗 www.b-s-h.org.uk/guidelines/

- Diagnosis and management of aplastic anaemia
- Primary autoimmune haemolytic anaemia
- Drug-induced immune and 2° autoimmune haemolytic anaemia

Patient information and support

Aplastic Anaemia Trust 🔗 www.theaat.org.uk

Haemoglobinopathy

Screening

- **Preconceptual screening** Consider in at-risk groups and when investigating or treating infertility
- **Antenatal haemoglobinopathy screening** (➔ p. 785) Offered to all pregnant women in the UK, ideally at <10wk gestation. If the mother is a carrier, the father is offered testing ± the fetus
- **Neonatal blood spot screening** (➔ p. 830) For sickle cell disease is offered to all babies born in the UK and those arriving before 1y

❗ Screening results in people knowing their carrier status. Ensure results are clearly recorded in patient notes. Issue haemoglobinopathy cards to those with haemoglobinopathy and who are confirmed carriers.

Thalassaemia Autosomal recessive inherited disorder of production of α (α-thalassaemia) or β (β-thalassaemia) globin chains of haemoglobin. Many varieties are recognized, but classified into 2 main types:

- α° and β° thalassaemia: no gene product is produced
- α^+ and β^+ thalassaemia: $\alpha + \beta$ chains present but produced at ↓ rate

β thalassaemia Occurs in all ethnic groups other than Northern Europeans. Defective β-chain production results in ↑ α-chain synthesis. Excess α chains precipitate in red cell precursors causing their destruction in bone marrow and spleen. This causes proliferation of marrow, bony deformity (mongoloid facies, bossing of skull, thinning of long bones), and progressive splenomegaly.

Homozygotes develop profound anaemia from 3mo of age and without repeated transfusions would die in <1y. If suspected refer urgently to paediatrics. ❗ Most infants with β° thalassaemia (β thalassaemia major) are detected at neonatal bloodspot screening.

Specialist ongoing care is essential. Offer family members referral for genetic counselling. Children who receive transfusions grow and develop normally but iron accumulates; chelation ↑ survival but iron overload may cause premature death. Stem cell transplant is curative.

α thalassaemia Occurs in populations from China, South East Asia, Greece, Turkey, Cyprus, and Sardinia.

- The homozygous state for α° thalassaemia is associated with fetal death at ~38wk (Barts hydrops)
- Haemoglobin H results from inheritance of α° from one parent and α^+ from the other; patients are moderately anaemic with splenomegaly and have haemoglobin H (4 β chains combined with a haem molecule) in their red cells. Specialist management is needed

Asymptomatic patients Heterozygotes for α or β thalassaemia (thalassaemia trait), and homozygotes for α^+ thalassaemia, are often asymptomatic. They may have mild anaemia with hypochromic/microcytic red cells. ❗ Can be confused with iron deficiency—suspect if MCV is disproportionately low; ferritin is normal; no response to iron.

Patient information and support

UK Thalassaemia Society ☎ 020 882 0011 🖱 www.ukts.org

The sickling disorders Inherited disorders most common among people originating from malarial areas—Africans (1–2% newborns) and certain Mediterranean, Middle Eastern, and Indian populations. *Varieties*:

- Heterozygous state for haemoglobin S (sickle cell trait—AS)
- Homozygous state (sickle cell anaemia/disease—SS)
- Heterozygous states for haemoglobin S and haemoglobins C, D, E or other structural variants
- Combination of haemoglobin S with any form of thalassaemia

Mechanism Haemoglobin S undergoes liquid crystal formation as it becomes deoxygenated causing sickling of affected blood cells. The effect of sickling is to shorten survival of red cells → haemolytic anaemia, and cause aggregation of the sickled cells, which in turn leads to:

- Tissue infarction—resulting in pain and/or tissue damage, e.g. stroke (10% children with sickle cell anaemia have a stroke) *and/or*
- Sequestration in the liver, spleen, or lungs—producing sudden and profound anaemia

Diagnosis FBC and film—chronic anaemia with sickling on film. Confirm diagnosis with haemoglobin electrophoresis.

Sickle cell trait Patients with <40% haemoglobin S have no symptoms unless they are subjected to anoxia, e.g. anaesthesia.

Sickle cell anaemia Low Hb level (typically 8–9g/dL) with high reticulocyte count—although generally patients compensate well. Illness results from complications arising from acute exacerbations or 'crises' and the effects of recurrent tissue damage due to microinfarction. Prognosis is variable. In Africa, children usually die in <1y. In the UK, patients survive into adulthood (average survival 50y). The most common cause of death is infection.

Patients should be managed by specialist centres, aiming to prevent crises (hydroxycarbamide may be used) and treat complications early. There is no medication to prevent sickling. *GPs should*:

- Treat patients as if hyposplenic—give HiB, meningitis C, pneumococcal + annual influenza vaccination, ± prophylactic antibiotics (➔ p. 626)
- Advise patients to avoid cold and maintain adequate hydration; warn about the dangers of anaesthetics (a MedicAlert bracelet is helpful)
- Treat infection early—be alert for aplastic crisis following parvovirus
- Give analgesia for painful crises (including weak opioid but strong opioids are ***not*** recommended for 1° care); if severe admit
- Admit if significant crisis of any sort (e.g. stroke, acute abdomen); have low threshold to admit children or if fever >38°C or chest symptoms
- Refer for early management of long-term complications (e.g. renal failure, epilepsy)

Patient information and support

Sickle Cell Society ☎ 020 8961 7795 🖱 www.sicklecellsociety.org

Further information

British Society for Haematology (2010) Significant haemoglobinopathies: guidelines for screening and diagnosis. 🖱 www.b-s-h.org.uk/guidelines

Bleeding and clotting disorders

Coagulation tests (sodium citrate tube; false results if under-filled)

- **Prothrombin time (PT)** Prolonged by coumarins (e.g. warfarin); vitamin K deficiency; liver disease
- **Activated partial thromboplastin time (APTT)** ↑ in heparin treatment, haemophilia, antiphospholipid syndrome, or DIC
- **INR** Ratio of the time the sample takes to clot compared to a control

The purpuras

Vascular purpuras Result from damage to the vessel wall. *Due to*:

- Infection (e.g. meningococcal septicaemia, EBV)
- Immune dysfunction (e.g. Henoch–Schönlein purpura)
- Vitamin C deficiency
- Ageing (senile purpura)
- Local stasis or ↑ venous pressure (e.g. varicose veins)
- Drug reaction (e.g. steroid-induced purpura)

Thrombocytopenic purpura Purpura is related to the level of the platelet count. Bleeding is inevitable if platelet count ↓ to <5–10 × 10^9/L.

- **Non-immune thrombocytopenic purpura** Results from conditions that damage the bone marrow, e.g. drugs (chemotherapy), aplastic anaemia (➲ p. 643), haematological malignancy (➲ p. 650)
- **Immune thrombocytopenic purpura** Usually primary immune (ITP) but may be 2° to SLE, transfusions, or drug reactions (e.g. heparin)

Idiopathic thrombocytopenic purpura (ITP)

- **In children** Self-limiting, often after a viral illness; platelet count is generally <10 × 10^9/L but severe bleeding is rare. Refer to paediatrics as an emergency. Usually no specific treatment is needed
- **In adults** Presents with haemorrhage and bruising. May be acutely symptomatic (sometimes recurrent), or chronic and insidious. Platelet count is ↓. Ask about drug history (particularly thiazides, quinine, or digoxin). Look for evidence of SLE or lymphoma. Examine for splenomegaly. Refer to haematology

Thrombotic thrombocytopenic purpura (TTP) Rare congenital or immune condition. May be rapidly fatal (90% mortality if untreated). Thrombocytopenia is accompanied by microangiopathic haemolytic anaemia, and small vessel thrombosis—usually causing neurological symptoms. If suspected, admit as medical emergency for plasma exchange.

Impaired platelet function May occur with myeloproliferative disorder/myelodysplasia and with very high paraproteins. Causes bleeding—even if platelet count is normal.

Haemophilia 2 common forms—haemophilia A (factor VIII deficiency) and B (factor IX deficiency—Christmas disease). Sex-linked recessive disorders but 1 in 3 results from a new mutation. ♂ >> ♀. *Prevalence*: 1/10,000 (haemophilia A); 1/50,000 (haemophilia B).

Classification:

- Carrier: ♀ heterozygotes; >25% clotting factor activity
- Mild: 5–25% clotting factor activity
- Moderate: 1–5% clotting factor activity
- Severe: 50% people with haemophilia; ≤1% clotting factor activity

Features Bleeding into joints/muscles—often delayed following trauma. Severity is related to levels of clotting factors. Untreated can cause permanent damage. Pressure effects occur if bleeding takes place into a confined space, e.g. intracranial bleed.

Management Follow-up should be via a specialist haemophilia centre. Prenatal/antenatal screening is available—refer to genetics.

- On-demand treatment Transfusion of factor VIII or IX preparation soon after bleeding has started—most self-administer; symptomatic treatment, e.g. rest, analgesia ± physiotherapy for muscle/joint bleeds
- Prophylaxis Prevents bleeds and their consequences. Agents used:
  - Tranexamic acid—prevents bleeding after minor surgical procedures for patients with mild haemophilia and carriers with symptoms
  - Desmopressin—stimulates production of factor VIII (not factor IX). Prevents/treats bleeding in mild/moderate haemophilia A
  - Regular factor VIII or factor IX for severe haemophilia

Problems with treatment 25% of patients have antibodies to factor VIII or IX products. Historically, blood products have resulted in HIV, and hepatitis B/C transmission. Not a risk with modern 'recombinant' products. ❗ All patients should have hepatitis B vaccination.

Patient information and support

Haemophilia Society ☎ 0207 9390780 🖱 www.haemophilia.org.uk

von Willebrand's disease Genetic—most autosomal dominant. ↓ levels of vW factor leads to menorrhagia, or ↑ bleeding after surgery (including dental procedures). Severity is linked to level of deficiency. Most bleeds are mild. *Prevalence*: 1% have low vW factor—many are asymptomatic. ♂ = ♀. *FBC*—usually normal platelets; *clotting screen*—often ↑ partial prothrombin time. Refer to haematology. Mild cases are managed with tranexamic acid, desmopressin, and/or COC or Mirena® for menorrhagia. Severe cases are treated with vW factor.

Thrombophilia^{G} ↑ tendency to clot. May be acquired or inherited:

Acquired 2° to obesity, immobility, pregnancy, COC, cancer, surgery (<6wk), smoking, antiphospholipid syndrome. Treat the cause if possible; otherwise discuss thromboprophylaxis with the relevant specialist. ❗ Patients <40y with unprovoked proximal DVT/PE or <50y with ischaemic stroke should be tested for antiphospholipid antibodies.

Inherited Defects in natural anticoagulants (e.g. protein S) and clotting factors (e.g. factor V Leiden). Refer/discuss with haematology if:

- Neonate/child with purpura fulminans (urgent testing)
- FH of 'high-risk thrombophilia' (protein C or S deficiency; antithrombin deficiency)
- Strong FH of unprovoked venous thrombosis (or for ♀, oestrogen-provoked thrombosis)
- Adults that develop skin necrosis on vitamin K antagonists

Further information

British Society for Haematology 🖱 www.b-s-h.org.uk/guidelines/

- Testing for heritable thrombophilia
- Thrombotic thrombocytopenic purpura
- Diagnosis and management of von Willebrand disease

Anticoagulation

Anticoagulants + antiplatelet therapy Except <12mo after stenting or acute coronary syndrome, antiplatelet agents are usually stopped if initiating anticoagulation. Avoid combining except on specialist advice (↑ risk of bleeding—clopidogrel > aspirin).

Heparin Enhances antithrombin III activity; sc LMWH is used in the community. Indications include:

- Initial treatment of VTE (while awaiting diagnostic testing and/or until oral anticoagulation is established)
- VTE prophylaxis in hospitalized/immobilized patients
- VTE management for cancer patients and pregnant women at high risk
- Bridging when warfarin is stopped prior to surgery
- High-risk patients on oral anticoagulants if INR is < desired range

Initiation of longer-term anticoagulation Options—coumarin anticoagulant (warfarin) or direct oral anticoagulant (DOAC). Consider risks/benefits of anticoagulation and pros/cons of DOAC/warfarin for each patient. Check baseline FBC, clotting screen, and renal/liver function.

Warfarin Antagonizes vitamin K to ↓ clotting tendency. Its effects can be reversed with vitamin K/prothrombin complex. Requires regular blood monitoring and dose adjustment. *Target INR:*

- Most indications 2.5 (range 2–3)
- Thromboembolism on anticoagulant treatment 3.5 (range 3–4)
- Prosthetic heart valves Take specialist advice

Indications

- Prevention of arterial thromboembolism in patients with cardiac disease, e.g. non-valvular AF (➔ p.240); some valvular heart disease, all mechanical prosthetic valves; dilated cardiomyopathy
- VTE—≥6wk after below knee DVT; ≥3mo after proximal DVT/PE; prevention of recurrent VTE (e.g. antiphospholipid syndrome)

Starting and monitoring Starting regimen—Table 18.4. Monitoring:

- 1 high INR—Recall 7–14d. Stop warfarin for 1–3d and restart at a lower dose. ⚠ If INR >8 and bleeding—admit; if not bleeding, consider admission or treat with vitamin K 2mg po or 1mg IV
- 1 low INR—↑ dose and recall in 7–14d
- If therapeutic INR: 1 therapeutic INR—recall in 4wk; 2 INRs—recall in 6wk (maximum interval if prosthetic heart valve); 3 INRs—recall in 8wk; 4 INRs—recall in 10wk; 5 therapeutic INRs—recall in 12wk

Patient advice

- Carry an anticoagulant card/booklet
- Take warfarin every evening
- Advise about local monitoring systems and/or self-monitoring
- If starting a drug that may interact with warfarin, recheck INR in 5–7d

Direct oral anticoagulants (DOACs) Agents which inhibit thrombin (e.g. dabigatran) or factor Xa (e.g. rivaroxaban, edoxaban, apixaban). Indications include:

- VTE—prophylaxis after orthopaedic surgery; treatment of VTE (initial and ongoing); prevention of recurrent VTE
- Stroke prevention in patients with non-valvular AF

Table 18.4 Dose regimen for starting warfarin in the community

INR on day 5	Dose on days 5–7	INR on day 8	Dose from day 8	Instructions
≤1.7	5mg	≤1.7	6mg	• Give warfarin 5mg od for 4d then check INR • Adjust dose as shown in table • Recheck INR on day 8 and adjust dose as shown in table • Thereafter check INR weekly (unless 4d interval stated) and adjust dose accordingly until dose is stable in the target range ⚠ **High INR** *INR ≥8* (lower if other risk factors for bleeding)—consider treating with oral/IM vitamin K or admit; always admit if bleeding *INR >3.7 and <8*—omit warfarin 1–2d and recheck INR. Restart when INR <5 and re-titrate dose
		1.8–2.4	5mg	
		2.5–3	4mg	
		>3	3mg for 4 d	
1.8–2.2	4mg	≤1.7	5mg	
		1.8–2.4	4mg	
		2.5–3	3.5mg	
		3.1–3.5	3mg for 4d	
		>3.5	2.5mg for 4d	
2.3–2.7	3mg	≤1.7	4mg	
		1.8–2.4	3.5mg	
		2.5–3	3mg	
		3.1–3.5	2.5mg for 4d	
		>3.5	2mg for 4d	
2.8–3.2	2mg	≤1.7	3mg	
		1.8–2.4	2.5mg	
		2.5–3	2mg	
		3.1–3.5	1.5mg for 4d	
		>3.5	1mg for 4d	
3.3–3.7	1mg	≤1.7	2mg	
		1.8–2.4	1.5mg	
		2.5–3	1mg	
		3.1–3.5	0.5mg for 4d	
		>3.5	Omit for 4d	
>3.7	0mg	<2	1.5mg for 4d	
		2–2.9	1mg for 4d	
		3–3.5	0.5mg for 4d	

Advantages of DOACs over warfarin

- Rapid onset of action
- Fixed dose
- No regular monitoring
- Fewer interactions

Disadvantages of DOACs

- Short half-life—good compliance is needed
- Not as readily reversible as warfarin—but as half-life is short, most bleeds can be managed with supportive treatment. New agents are in development to reverse the action of DOACs
- May not be suitable if impaired renal function—check/record creatinine clearance regularly (>1×/y) and consult product literature

Bleeding on anticoagulants HAS-BLED score—Table 15.9, ➔ p. 537. Risk of major bleed/death are similar for DOACs and warfarin:

- Major bleeding—admit
- Head injury—if laceration, bruising, LOC, amnesia, persistent headache, or ↓ GCS—refer to A&E for CT scan
- Bleeding on DOAC or warfarin at therapeutic INR (e.g. haematuria, rectal bleeding)—investigate cause

Haematological malignancy

⚠ **Suspected haematological malignancy**^{N} May present with non-specific symptoms/signs. Have a high level of suspicion.

Immediate specialist assessment (haematology or paediatrics)
- Children/young people—unexplained petechiae or hepatosplenomegaly
- FBC/blood film reported as showing acute leukaemia
- Suspected spinal cord compression
- Suspected renal failure associated with myeloma

Specialist assessment in <48h for children/young people and in <2wk for adults Consider if unexplained lymphadenopathy or splenomegaly. Take associated symptoms into account, e.g. fever, night sweats, shortness of breath, pruritus, weight ↓.

Specialist assessment in <2wk if
- Protein electrophoresis or a Bence Jones protein urine test suggests myeloma

FBC in <48h
- Pallor
- Unexplained fever
- Generalized lymphadenopathy
- Unexplained bruising/bleeding
- Hepatosplenomegaly in adults
- Unexplained petechiae in adults
- Persistent fatigue
- Unexplained persistent infection (or recurrent infection in adults)
- Persistent or unexplained bone pain in children

Protein electrophoresis + Bence Jones protein in <48h if
- >60y + hypercalcaemia/leucopenia + symptoms/signs of myeloma
- Plasma viscosity/ESR + presentation consistent with myeloma

FBC, Ca^{2+}, and plasma viscosity/ESR If ≥60y + persistent bone pain (particularly back pain), or unexplained fracture.

Multiple myeloma Age usually >60y. 5500 new cases/y in the UK and 2900 deaths. A mutant plasma cell clone is present. The proliferating cells grow mainly in the bone marrow where they cause infiltration, localized tumours, and bone erosion. Main sites of myeloma involvement are skull, spinal column, thoracic cage, pelvis, and proximal long bones.

Presentation
- Infection, e.g. chest infection
- Anaemia and/or bleeding
- Bone pain ± tenderness—particularly ribs, back, pelvis
- Pathological fracture
- Hypercalcaemia
- Renal failure
- Hyperviscosity syndrome (CNS features, e.g. blurred vision, altered consciousness, confusion)
- Amyloidosis (heart, tongue, carpal tunnel)

Investigation

- FBC—anaemia; blood film—rouleaux formation
- Renal function ↑ Cr; ↓ eGFR
- ESR ↑↑
- Ca^{2+} frequently ↑
- Serum electrophoresis Paraprotein band
- Urine electrophoresis Bence Jones protein (BJP)—useful for diagnosis (❗ occasionally may be non-secretory when BJP will be −ve)
- Serum free light chains (blood equivalent of BJP) Used to measure response to treatment
- X-ray Erosive lesions in skull, ribs, pelvis. Fractures and vertebral collapse are common

Management Refer urgently to haematology. Specialist management depends on symptoms and whether there is tissue/organ damage.

- Asymptomatic disease Patients are usually monitored closely and treatment starts if symptoms or tissue/organ damage develop
- Symptomatic disease Treatment options include chemotherapy, steroids, biological agents (bortezomib), and/or novel agents (thalidomide, lenalidomide) as well as maintenance therapy. Intensive chemotherapy + stem cell transplant is offered to younger patients. Supportive treatment includes transfusions; management of infections: thromboprophylaxis; bisphosphonates for hypercalcaemia and bone pain (bone pain may also respond to radiotherapy); dialysis for renal failure; and plasmapheresis for hyperviscosity
- Relapsed disease Patients who have gone into remission almost always relapse at some point. Novel/biological agents are frequently used in the treatment of relapse

Prognosis Survival is improving and ranges from weeks to many years; overall 10y survival is 33% but higher in men and those diagnosed at a younger age.

Monoclonal gammopathy of undetermined significance (MGUS) Presence of monoclonal paraprotein band in isolation with no other features of myeloma or other lymphoproliferative disease. Present in 1% >50y and 5% if >80y. Usually found incidentally. Most remain stable but ~1%/y progress to myeloma or other haematological malignancy.

Management

- Exclude myeloma, other lymphoproliferative disorders, and amyloidosis; refer urgently if haematological malignancy/amyloidosis is suspected
- Refer to haematology if IgD or IgE M-protein, >15g/L IgG M-protein, or >10g/L IgA or IgM M-protein
- Monitor clinical symptoms and check M-protein (↑ >25% should prompt re-referral), immunoglobulins, FBC, ESR, U&E, Cr, and eGFR every 3–4mo for the first year then every 6–12mo; re-refer if any significant, sustained change
- Advise patients to re-attend promptly if new symptoms (e.g. back pain, fatigue, weight ↓) develop

Further information

British Society for Haematology (2009) Investigation of newly detected M-proteins and the management of MGUS. 🖱 www.b-s-h.org.uk/guidelines

NICE (2017) Suspected cancer: recognition and referral. 🖱 https://www.nice.org.uk/guidance/ng12

Acute leukaemia

Clonal malignant disorders (from a single cell) affecting all age groups.

Acute lymphoblastic leukaemia (ALL) Abnormal proliferation in the lymphoid progenitor cells (Figure 18.2). Incidence is 1–4/100,000 population/y. ♂ > ♀. *Usual age range*: 2–10y with a peak at 3–4y. Accounts for 85% of childhood leukaemia. Incidence then falls with increasing age apart from a secondary peak at ~40y.

Acute myeloid leukaemia (AML) Abnormal proliferation of a myeloid progenitor cell (Figure 18.2). There are several different subtypes. Most common leukaemia of adulthood with incidence of ~1.5/100,000 population/y. Incidence ↑ with age. Median age at presentation ≈60y. ♂ = ♀. Risk factors: smoking (1 in 5 cases); previous chemotherapy/radiotherapy; exposure to radiation; chemical exposure (e.g. benzene); certain genetic syndromes, e.g. Down's syndrome; myeloproliferative disorders.

Presentation Short history (weeks). Symptoms/signs arise from:

Bone marrow failure Anaemia—pallor, lethargy, dyspnoea; neutropenia—infections of the mouth, throat, skin, fever; thrombocytopenia—spontaneous bruising, menorrhagia, bleeding from wounds, bleeding of gums or nose bleeds.

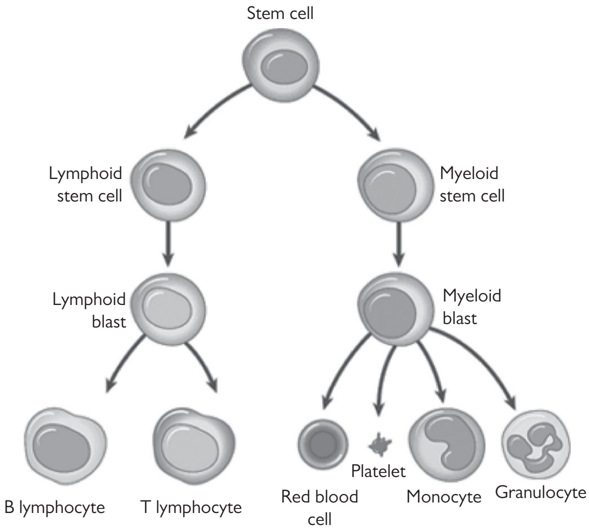

Stem cell
Lymphoid stem cell
Myeloid stem cell
Lymphoid blast
Myeloid blast
B lymphocyte
T lymphocyte
Red blood cell
Platelet
Monocyte
Granulocyte

Figure 18.2 Blood cell production

Taken from CancerHelp UK, the patient information website of Cancer Research UK: http://www.cancerresearchuk.org/cancerhelp

Table 18.5 Prognosis of acute leukaemia

	Overall 5y survival
Childhood ALL	88%
Adult ALL	40% (80% achieve remission)
AML age <55y	40% (>60% for children)
AML age >55y	<20%

Organ infiltration Superficial lymphadenopathy (>50%); hepatosplenomegaly (70%); bone pain (ALL only); skin infiltration (AML only); testicular enlargement; respiratory symptoms 2° to mediastinal LNs; gum hypertrophy; unexplained irritability/behaviour change/↓ performance.

Differential diagnosis Infections, e.g. EBV; other blood conditions, e.g. aplastic anaemia, ITP, myelodysplasia; other malignancies, e.g. lymphoma, neuroblastoma, metastatic disease; rheumatoid arthritis.

Investigation

- FBC Hb and platelets normal or ↓; WCC $<1 \times 10^9$/L to $>200 \times 10^9$/L
- Blood film Abnormal with presence of blast cells
- Renal function Renal impairment if leucocyte count is very high
- CXR May show mediastinal mass and/or lytic bone lesions

Initial management^{N} Refer for same-day specialist opinion if:

- Abnormal blood count reported as needing urgent investigation
- Unexplained petechiae in a child
- Hepatosplenomegaly in a child

Specialist management Once diagnosis is confirmed, treatment is co-ordinated in specialized centres and involves intensive supportive care together with systemic chemotherapy, ± radiotherapy ± stem cell transplant. For patients with ALL, treatment includes maintenance therapy for 2y to help maintain remission. Prognosis—Table 18.5.

Short-term side effects of treatment

- Treatment side effects Most chemotherapeutic agents have pronounced side effects, e.g. nausea, vomiting, hair loss, neuropathy
- Immunosuppression Any neutropenic child or adult presenting with fever must be taken seriously and referred immediately back to the unit in charge of care. Any chickenpox contact must be referred immediately for consideration of administration of varicella zoster Ig, or measles contact for administration of measles Ig

Long-term side effects of treatment Heart—cardiomyopathy, arrhythmias; lung—fibrosis; endocrine system—growth delay, hypothyroidism, infertility; kidney—↓ eGFR; 2° malignancies—may appear after many years; psychological effects.

Information and support for patients and carers

Bloodwise ☎ 0808 2080 888 www.bloodwise.org.uk
CLIC Sargent ☎ 0300 330 0803 www.clicsargent.org.uk
Macmillan Cancer Support ☎ 0808 808 0000 www.macmillan.org.uk

Chronic leukaemia and myeloproliferation

Chronic lymphocytic leukaemia (CLL) Mainly affects the elderly accounting for 40% of leukaemias in that age group. Closely related to small lymphocytic lymphoma (Table 18.6, ➔ p. 657). >80% of diagnoses follow FBC done for another reason; many never develop symptoms or signs. Otherwise presents with widespread painless lymphadenopathy often noted over a period of months/years, weight ↓ and sweats may occur. *Examination*: check for lymphadenopathy, spleno- ± hepatomegaly.

Investigation ↑ lymphocyte count ($>5 \times 10^9$/L). Can be diagnosed from a peripheral blood film (small lymphocytes—disrupted to form ‘smear’ cells) and immunophenotyping to identify clonal B cells.

Management Asymptomatic patients with low levels of lymphocytosis can be managed in 1° care:

- Monitor FBC and perform clinical review initially 6-monthly
- Manage infection risk by offering influenza and pneumococcal vaccination, but NOT shingles vaccine. Treat infections promptly. Advise about ↑ risk of other malignancies including skin cancer

Refer To haematology if:

- Symptomatic disease (recurrent infections, fevers, sweats, weight ↓)
- Bulky lymphadenopathy and/or hepatosplenomegaly
- Anaemia or thrombocytopenia
- Rising lymphocyte count (↑ >50% in 2mo or doubling time of <6mo)

Specialist treatment Chemotherapy, monoclonal antibody therapy, and, in a few cases, stem cell transplant.

❗ The term ‘leukaemia’ provokes fear in many. Explain the diagnosis of CLL, its benign nature in many, and that prognosis can be >10y.

Chronic myeloid leukaemia (CML) Rare in children, median age of presentation is 65y. Chance finding in >20%. Otherwise presents with:

- Non-specific symptoms, e.g. weight ↓, lassitude, gout, anaemia
- Splenomegaly (common)—abdominal pain, digestive symptoms, or pleuritic pain due to splenic infarction
- Bleeding (rare)—due to abnormal platelet function

Investigation FBC: ↑ WCC (usually $>50 \times 10^9$/L) ± anaemia. *Blood film*: bone marrow precursors of myeloid cells (blasts). *Cytogenetics*: Philadelphia chromosome.

Management Refer urgently to haematology. Treatment is determined by phase of the disease:

- Chronic (90% at diagnosis) <10% of cells in the bone marrow are immature blasts. Treatment with tyrosine kinase inhibitors (e.g. imatinib) has dramatically improved prognosis in recent years
- Accelerated 10–30% of cells in the bone marrow are immature blasts. Treatment is with chemotherapy ± stem cell transplant, or 2nd-generation tyrosine kinase inhibitors
- Blast (also called acute phase, blast crisis) >30% of cells in the bone marrow are immature blasts—treated with chemotherapy

Myeloproliferative disorders Proliferation of ≥1 of the haemopoietic components of the bone marrow. Includes:

- CML
- Polycythaemia vera
- Essential thrombocythaemia
- Myelofibrosis

Erythrocytosis ↑ in the number of circulating red cells. If haematocrit is persistently ↑ for >2mo (>0.52 ♂; >0.48 ♀), investigate the cause. May be 1° (polycythaemia vera) or 2°. ❗ Hb may also appear ↑ if dehydrated (concentration effect). 2° polycythaemia may be:

- Appropriate High altitude, chronic lung disease (e.g. COPD), cardiovascular disease with a right → left shunt, heavy smoking, sleep apnoea, ↑ affinity for haemoglobin (familial polycythaemia) *or*
- Inappropriate Caused by excess erythropoietin, e.g. 2° to hepatocellular or renal tumour, or massive uterine fibroid. May need venesection

Polycythaemia vera Also known as primary proliferative polycythaemia (PPP). Haematological malignancy resulting in overproduction of red cells. *Age range*: most >50y. Presentation is non-specific with:

- Night sweats
- Dusky, cyanotic hue with red face
- Itching (especially provoked by water, e.g. after a bath)
- Splenomegaly (70%) ± hepatomegaly
- Headaches, dizziness, vertigo, and/or tinnitus.
- Gout—2° to ↑ red cell turnover
- Peptic ulceration (5–10%)
- Thrombosis/haemorrhage—abnormal platelet function/hyperviscosity

Investigation Often diagnosed incidentally following FBC done for other reasons—persistently ↑ Hb + haematocrit; neutrophils and platelets may also be ↑. JAK2 mutation is +ve in >95%.

Management Refer urgently to haematology. Hb level is ↓ by regular venesection ± cytotoxics. Aspirin ↓ risk of thrombosis. Slowly progressive and survival for 10–20y is not unusual; a minority eventually transform to AML or myelofibrosis.

Essential thrombocythaemia Patients have ↑ risk of thrombosis, but may haemorrhage due to abnormal platelet function. *FBC*—persistently ↑ platelet count >450 × 10^9/L when reactive (including iron deficiency) and other myeloproliferative causes have been excluded. Refer urgently to haematology. Treatment is with aspirin, treatment of CVD risk factors, ± cytotoxics. May transform to AML or myelofibrosis.

Myelofibrosis (myelosclerosis) Progressive accumulation of fibrous tissue in the bone marrow cavity replacing normal marrow. Haemopoietic function is taken over by the spleen/liver. Patients are usually elderly and present with symptoms of anaemia, malaise, fever, ± gout. The spleen is massively enlarged. *FBC*—↓ Hb; *Blood film*—immature erythroid cells (normoblasts) and myeloid cells (metamyelocytes/myelocytes). Red cells are tear-drop shaped. Refer urgently to haematology. 5y survival is 48%—many live much longer. 5–10% transform to AML.

Further information

British Society for Haematology www.b-s-h.org.uk/guidelines

- Thrombocytosis
- Myelofibrosis
- Polycythaemia/erythrocytosis
- CLL

Lymphoma

Cancer of the lymphatic system. 2 main types.

Non-Hodgkin's lymphoma (NHL) Derived from malignant transformation of lymphocytes—85% B cells. Usually develops in LNs but can arise in any tissue. *Incidence*: 13,600 cases/y in the UK (4% cancers), causing 4800 deaths/y. ♂ = ♀. 49% occur in patients >70y.

Presentation May be detected incidentally on CXR (mediastinal mass) or present with painless peripheral lymphadenopathy; abdominal mass (nodal or spleen); weight ↓; night sweats/unexplained fevers. Other symptoms are dependent on site, e.g. neurological symptoms if CNS involvement; pleural effusion; skin lesions. See Table 18.6.

Investigation *FBC*—may be normal if no bone marrow involvement; *monospot*—for all patients <30y with persistent lymphadenopathy to exclude EBV; *ESR*—usually ↑; *LFTs*—abnormal if liver involvement.

Initial management Depending on local referral pathways, consider urgent referral to oncology/haematology if:

- Unexplained lymphadenopathy[N]—➔ p. 916
- LNs are ↑ in size or LN >2cm in size
- Widespread lymphadenopathy
- Lymphadenopathy + weight ↓, night sweats, and/or splenomegaly
- Any other suspicious symptoms/signs

Specialist treatment Based on histology and stage. Treatment options include a watchful waiting approach for low-grade lymphomas; radiotherapy; chemotherapy; stem cell transplant; monoclonal antibody therapy (rituximab); and/or immunotherapy.

Prognosis Varies widely between different types of NHL and the age of the patient. Younger, fitter patients with less widespread disease do better. Overall survival improving to 63% at 10y.

Hodgkin's lymphoma 2100 cases/y in the UK. *Peak age ranges*: 15–35y (50% occur <45y) and 65–85y. Derived from B lymphocytes. 2 types of Hodgkin's lymphoma are recognized:

- Classical (95%) Reed–Sternberg cells are present
- Nodular lymphocyte predominant (5%) 'Popcorn' cells are present

Presentation Painless lymphadenopathy (70–95% have affected cervical LNs at diagnosis), weight ↓, night sweats/unexplained fevers, pruritus. The spleen is involved in 30% → splenomegaly.

Investigation and management As for NHL.

Prognosis

- Early stage disease (Ann Arbor stage 1/2—affected lymph tissue is confined to 1 side of the diaphragm)—90% 5y survival
- Late stage disease (Ann Arbor stage 3/4—affected lymph tissue both sides of diaphragm and/or extralymphatic tissue involvement)—75–90% 5y survival

Information and support

Lymphoma Association ☎ 0808 808 5555 🖱 www.lymphomas.org.uk

Table 18.6 Features of common types of NHL

Type	Features
High-grade NHL	
Diffuse large B cell (DLBCL)	48% NHL (including childhood). Median age at diagnosis 70y Presents with rapidly enlarging lymphadenopathy. Extranodal involvement is common. 10% have bone marrow involvement at presentation. 5y survival 60%
Anaplastic large cell	2 forms. Both originate from T cells or unknown cells *Systemic form*: affects children/young adults. ♀ > ♂. Usually presents at a late stage and with systemic symptoms *Cutaneous form*: affects adults. Presents with reddish brown skin nodules or ulceration ± regional LN involvement (25%)
Burkitt's lymphoma	30–40% childhood lymphoma. ♂ > ♀. B-cell lymphoma. 2 varieties. Endemic variety is more common in Africa and associated with EBV infection. Peak age 5–10y. Sporadic variety occurs worldwide and affects children and adults. 5y survival 56% Presents with bulky central nodal disease ± extranodal (typically abdomen), bone marrow, and/or CNS involvement
Low-grade NHL	
Follicular	19% NHL. Affects adults, median age at diagnosis 65y. B-cell origin 85% present with disseminated disease and 50% with bone marrow involvement. Indolent course but 20–30% transform to DLBCL. Prognosis varies with grade, stage, and age. Overall 5y survival 87%
Small lymphocytic	4–5% NHL. Median age 60y. Clinically/morphologically identical to CLL (➔ p. 654). Distinguished by degree of lymph tissue vs blood/bone marrow involvement Presents with diffuse lymphadenopathy and some blood/bone marrow involvement. 10–20% transform to CLL, 3% to DLBCL
Mantle cell	5% NHL. Affects adults usually >50y. ♂ > ♀ (4:1). Although classified as low grade, behaves and is treated as high grade. Usually presents with widespread disease involving LNs, bone marrow (60–90%), peripheral blood, spleen, ± gut. 5y survival 40%
Marginal zone	B-cell origin. 3 distinct types *Nodal*: 1–3% NHL. Presents with localized lymphadenopathy *Splenic*: <1% NHL. Affects adults. Presents with massive splenomegaly and blood/bone marrow involvement without lymphadenopathy *Mucosa associated (MALT)*: 10% NHL. May be associated with inflammation (e.g. *H. pylori* infection and gastric MALT; Hashimoto's thyroiditis and thyroid MALT). 70% have localized disease on presentation. Symptoms depend on the organ involved
Lympho-plasmacytic	1.5% NHL. Also called Waldenström's macroglobulinaemia. B-cell lymphoma. Average age at presentation is 63y Often presents late with lymphadenopathy, splenomegaly, and bone marrow involvement. May spread to the lung or GI tract. Usually associated with paraproteinaemia (IgM)

Immune deficiency syndromes

⚠ Consider an immunodeficiency disorder in anyone with infections that are unusually frequent, severe, resistant, or due to unusual organisms.

A group of diverse conditions caused by immune system defects and characterized clinically by ↑ susceptibility to infections.

History In addition to history of infection, ask about:

- Family history: immune deficiency, early death, similar disease, autoimmune illness, early malignancy
- Late separation of umbilical cord (>4wk) or shedding of 1° teeth
- Failure to thrive
- Adverse reaction to immunization or viral infection
- Difficult-to-treat asthma or eczema
- Splenectomy, tonsillectomy, or adenoidectomy
- Prior prophylactic antibiotic or immunoglobulin therapy

Primary immunodeficiency As many 1° immunodeficiencies are hereditary or congenital, they appear initially in infants and children; ~80% of those affected are <20y old and, due to X-linked inheritance, ♂ >> ♀. Genetic screening is available for some conditions.

Classification >70 1° immunodeficiencies are described They are classified into 4 groups depending on which component of the immune system is deficient (B cells, T cells, phagocytes, and complement).

Prevalence Selective IgA deficiency (usually asymptomatic) occurs in 1:400 people. All other 1° immune deficiencies are rare. Excluding IgA deficiency, 50% of affected patients have B-cell deficiency; 30% T-cell deficiency; 18% phagocytic deficiencies; and 2% complement deficiency.

Presentation Table 18.7 lists some of the more common immune deficiencies. All immune deficiencies present with ↑ tendency to infection. If suspected, refer for specialist review to paediatrics/immunology.

Secondary immunodeficiency Impairment of the immune system resulting from illness (including drug therapy, e.g. with cytotoxics or steroids) or removal of the spleen in a previously normal person. Often reversible if the underlying condition or illness resolves. 2° immunodeficiencies are common; most prolonged serious illness interferes with the immune system to some degree. Treat the cause.

HIV infection ➔ p. 720 **Asplenia** ➔ p. 626

Infection in the immunocompromised ➔ p. 626

Information and support for patients

Primary Immunodeficiency UK ☎ 0808 987 8986 🖱 www.piduk.org

Table 18.7 Immune deficiency syndromes

Type	Syndrome	Clinical details
B-cell deficiency Prone to infection with Gram +ve organisms (e.g. streptococci)	Selective IgA deficiency	May be associated with allergy and autoimmune disease and sometimes infections
	IgG subclass deficiencies	Most only mildly affected/asymptomatic, if more severely affected, early treatment of infection may be required. Usually improves with age
	Congenital X-linked hypogammaglobulinaemia and common variable immunodeficiency	↓ immunoglobulins Treatment is with IV immunoglobulin, antibiotics are needed for breakthrough infections and sometimes prophylactically ↑ risk of leukaemia/lymphoma/gastric cancer
T-cell deficiency Prone to viral, fungal, and opportunistic infections	DiGeorge's syndrome	Autosomal dominant, deletion on chromosome 22 → absent/hypoplastic thymus (and ↓ T cells), absent parathyroid glands, ± cardiac and/or facial abnormalities and developmental delay • Mild (80%)—treated supportively • Severe—requires thymus/stem cell transplant
	HIV	➔ pp. 720–3
Combined B- and T-cell deficiency	Severe combined immunodeficiency	Autosomal or X-linked recessive Absence of both T-cell and B-cell immunity Presents <6mo old with frequent infections Treatment is with bone marrow transplant Untreated most die at <1y
	Ataxia telangiectasia	Autosomal recessive Selective IgA deficiency or hypogammaglobulinaemia and T-cell dysfunction Characterized by telangiectasia, cerebellar ataxia, and recurrent chest infections Treatment is supportive ↑ risk of leukaemia/lymphoma
	Wiskott–Aldrich syndrome (partial combined immunodeficiency syndrome)	X-linked recessive ↑ IgA and IgE; normal or ↓ IgG; ↓ IgM Presents with eczema, thrombocytopenia, and recurrent infections Treatment is with stem cell transplant—rarely survive beyond teens without ↑ risk of leukaemia/lymphoma
Phagocytic deficiency Prone to staphylococcal and Gram −ve infections	Chronic granulomatous disease	X-linked (2/3) or autosomal recessive Phagocyte dysfunction Usually presents at <6mo of age with fungal pneumonia, lymphadenopathy, hepatosplenomegaly, and/or osteomyelitis Treatment is with prophylactic antibiotics/early treatment of infections, or stem cell transplant.
	Agranulocytosis	Usually caused by drugs, e.g. carbimazole Absence of neutrophils Sudden onset of fever ± rigors, sore throat, mouth ulcers, headache, and malaise → septicaemia If suspected, check urgent FBC and/or admit

Allergies

'One man's meat is another man's poison.'

Allergic diseases result from an exaggerated response of the immune system to external substances. Affect 1 in 6 of the British population—and is ↑. Allergic problems include:

- Asthma—➔ p. 278
- Occupational asthma—➔ p. 306
- Eczema—➔ p. 578
- Anaphylaxis—➔ p. 1054
- Drug allergy—➔ p. 120
- Urticaria—➔ p. 586
- Rhinitis—➔ p. 920
- Conjunctivitis—➔ p. 944
- Food intolerance
- Eosinophilic oesophagitis—➔ p. 356

Assessment

- Age
- Symptoms—past and present, main problem, frequency and severity, seasonal/perennial, provoking factors
- Impact on lifestyle—time off work/school, sleep
- Occupation/hobbies
- Treatment—past and present
- Home environment—pets, damp, dust, smoking
- Allergies in the past and/or family history of allergic illness
- Examination will depend on main symptoms (e.g. asthma—➔ p. 278)

Investigation

IgE skin prick and serum testing Identifies IgE sensitivity to common allergens, allowing diagnosis or exclusion of atopy. Uses skin prick testing or measurement of serum IgE levels. In most places this is a 2° care procedure although it is feasible in general practice. Patients should avoid using antihistamines before skin prick testing.

Patch testing Specialist test. Identifies substances causing contact allergy. A battery of allergens on discs are applied to the skin—usually on the back—and stuck in place with tape. Skin response is then monitored.

Management

Allergen avoidance For patients with anaphylaxis, may be lifesaving:

- **Pets** Exclude the offending animal
- **Pollens** Keep windows shut (including car windows); wear glasses/sunglasses; avoid grassy spaces; fit a pollen filter on the car
- **Foods/drugs** Avoid the food/drug; avoid hidden exposure (check labels carefully); inform any school/clubs a child attends; take food with you wherever possible; record drug allergies in medical notes

House dust mite Evidence that anti-house dust mite measures are effective in the relief of asthma and eczema is weak. Measures focus on the bedroom. Advise that the room should be ventilated regularly; encase mattresses, pillows, and duvets in mite-proof covers (leave in place 6mo); wash bed clothes at 60°C every 1–2wk.; use a vacuum cleaner with an adequate filter; remove bedroom carpet; ↓ soft toys to a minimum and wash frequently/put in the freezer to kill house dust mites.

Medication See individual conditions.

Referral to specialist allergy clinic

- Anaphylaxis
- Food allergy
- Occupational allergy
- If allergy diagnosis is in doubt
- Urticaria in which allergic aetiology is suspected
- For consideration of immunotherapy

Bee/wasp sting allergy Accounts for ~4 deaths/y in the UK. Stings may result in a local or generalized reaction of varying severity. Treat local or mild generalized reactions with antihistamine. Supply patients with more severe reactions with an adrenaline autoinjector pen and teach them, and close contacts, how to use it. If severe reaction, refer to an allergy clinic for consideration of desensitization.

Food allergy Affects 1.4% of the adult population and 5–7% of children. Types of adverse reaction to foods include:

- IgE-mediated food allergy Acute reaction (e.g. acute peanut allergy). Symptoms include itching, erythema, urticaria, angio-oedema, GI (e.g. vomiting, abdominal pain), sneezing, wheeze, anaphylaxis
- Non-IgE-mediated food allergy Delayed reaction (e.g. cow's milk causing eczema, eosinophilic oesophagitis). Symptoms include erythema, itching, eczema, persistent GI symptoms (e.g. reflux, pain, loose stools, constipation)
- Non-allergic food intolerance May be pharmacological, e.g. tyramine in red wine/cheese causing migraine; metabolic, e.g. lactase deficiency; or toxic, e.g. reaction to preservative rather than food
- Food aversion Symptoms are non-specific and unconfirmed by blinded food challenge

A limited number of foods are responsible for the majority of true food allergies: nuts (especially peanuts), wheat, eggs, fish, shellfish, cow's milk.

Management

- IgE-mediated reaction Request skin prick testing or send blood for specific IgE testing. Advise allergen avoidance and supply an adrenaline autoinjector pen (and teach how to use) if anaphylactic reaction
- Otherwise Try eliminating the suspected food for 2–6wk and then reintroduce. Dietician advice may be helpful

Referral Consider referral to allergy clinic or paediatrics if:

- Severe acute allergic reaction (e.g. anaphylaxis or angio-oedema)
- Severe delayed reaction (e.g. severe eczema related to food)
- Confirmed IgE allergy and asthma
- Suspected multiple allergies
- Poor growth with GI symptoms
- Parental concern
- Strong suspicion of allergy despite negative tests
- No response to food avoidance

Further information

NICE (2011) Food allergy in under 19s. www.nice.org.uk/guidance/cg116

Patient information and support

Allergy UK ☎ 01322 619898 www.allergyuk.org
Anaphylaxis Campaign ☎ 01252 542029 www.anaphylaxis.org.uk
MedicAlert Foundation ☎ 01908 951045 www.medicalert.org.uk
Medi-Tag ☎ 0121 200 1616 www.medi-tag.co.uk

Breast awareness means knowing what your breasts look and feel like normally. Evidence suggests that there is no need to follow a specific or detailed routine such as breast self-examination, but you should be aware of any changes in your breasts.

The breast awareness 5-point code

1. Know what is normal for you
2. Know what changes to look and feel for
3. Look and feel
4. Report any changes to your GP without delay
5. Attend for routine breast screening if you are aged 50y or over

Changes to be aware of

- Size If one breast becomes larger, or lower
- Nipples If a nipple becomes inverted (pulled in) or changes position or shape
- Rashes Or crusting on or around the nipple
- Discharge From one or both nipples
- Skin changes Puckering or dimpling
- Colour Redness or inflammation
- Lump or thickening Different to the rest of the breast tissue.

What should I do if I notice a change? If you do notice a change in your breasts, see your GP as soon as you can. Your GP may ask you to come back at a different time in your menstrual cycle, or send you to a breast clinic for a more detailed examination.

❗ Remember that most breast changes are not cancer, even if they need follow-up treatment or further investigation.

Further information

Breast Cancer Now 🖱 http://breastcancernow.org/sites/default/files/public/tlc_breast_awareness_guide.pdf
NHS Cancer Screening 'Be breast aware' leaflet. 🖱 www.gov.uk/government/uploads/system/uploads/attachment_data/file/439602/breastaware.pdf

Breast awareness 5-point code reproduced from NHS England. How should I check my breasts? © Crown copyright. Available at https://www.nhs.uk/common-health-questions/womens-health/how-should-i-check-my-breasts/. Contains public sector information licensed under the Open Government Licence v3.0.

Chapter 19

Breast disease

Breast symptoms *664*
Benign breast disease *668*
Breast cancer screening *670*
Risk factors for breast cancer *672*
Breast cancer: diagnosis and care *674*

Breast symptoms

⚠ **Urgent referral of patients with breast disease** (to be seen in <2wk by a breast cancer specialist) is required for men and women:

- Aged ≥30y with any unexplained breast lump ± pain *or* unexplained lump in the axilla
- Aged ≥50y with any of the following symptoms in 1 nipple only:
  - Discharge
  - Retraction
  - Other changes of concern
- Skin changes that suggest breast cancer, e.g. unilateral nipple eczema that does not respond to topical treatment, peau d'orange

Consider non-urgent referral if

- <30y with an unexplained breast lump ± pain
- Breast pain and no palpable abnormality, when initial treatment fails and/or symptoms persist

If DVT consider breast cancer as a cause. Carry out an assessment for additional symptoms, signs, or findings that may help to clarify whether breast cancer is the cause and refer urgently if breast cancer is suspected.

⚠ In patients presenting with symptoms and/or signs suggestive of breast cancer, investigation prior to referral is not recommended. Consider discussion with a specialist (e.g. by telephone or email) if there is uncertainty about the interpretation of symptoms/signs to decide whether a referral is needed.

Breast lump

History Age (malignancy is rare <30y); how and when noticed; relationship to menstrual cycle; changes in shape or size since noticed; pain; nipple discharge; skin changes; pregnancy and breastfeeding; family history; current medication (in particular contraceptive pill or HRT).

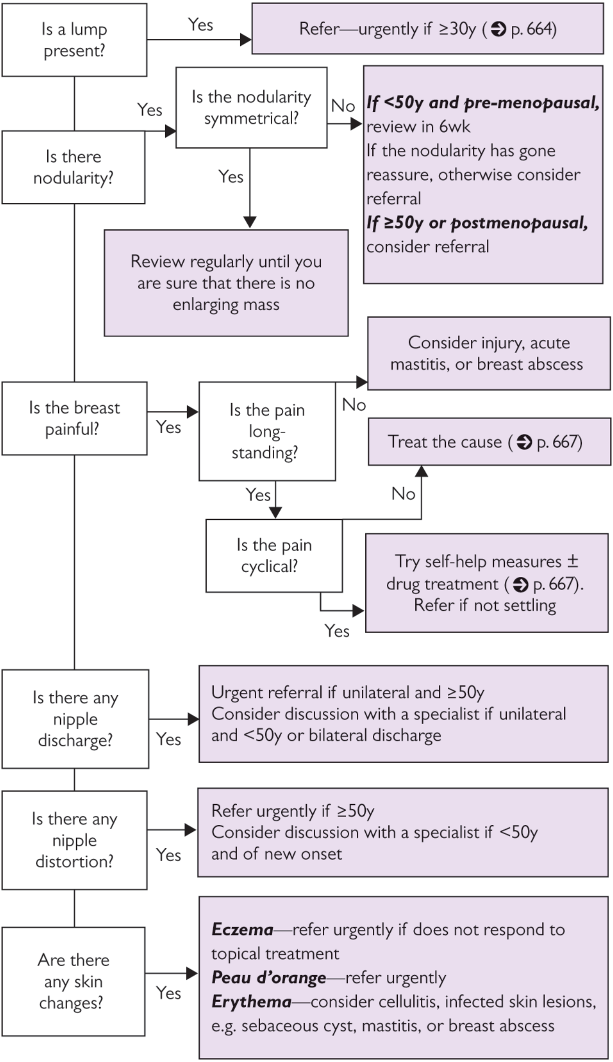
Examination See Box 19.1. Examine both breasts. If a lump is found, assess shape, size, surface, edge, consistency, mobility, and attachments. Check local LNs in axilla/supraclavicular region and for hepatomegaly.

Differential diagnosis Figure 19.1

- Breast cancer—➔ p. 674
- Fibroadenoma—➔ p. 668
- Breast cyst—➔ p. 668
- Duct ectasia/periductal mastitis—➔ p. 669
- Haematoma or fat necrosis—➔ p. 668
- Phyllodes tumour—➔ p. 668
- Intraductal papilloma—➔ p. 669
- Lipoma or sebaceous cyst

Box 19.1 Breast examination tips

- With the woman seated, with arms at her sides, above her head, and pressing on her hips, look at the size and shape of the breasts, skin contour, skin, and nipple changes
- Seat the woman at 45° supported on a couch. Ask her to place the hand on the side being examined behind her head. Ask the woman to point to or find the lump. Palpate each quadrant of the breast with a flat hand. Check the tail of the breast in the axilla


Is a lump present?
Yes
Is there nodularity?
Yes
Is the nodularity symmetrical?
No
Yes
Is the breast painful?
Yes
Is the pain long-standing?
No
Yes
Is the pain cyclical?
No
Yes
Is there any nipple discharge?
Yes
Is there any nipple distortion?
Yes
Are there any skin changes?
Yes

Refer—urgently if ≥30y (➔ p. 664)

If <50y and pre-menopausal, review in 6wk
If the nodularity has gone reassure, otherwise consider referral
If ≥50y or postmenopausal, consider referral

Review regularly until you are sure that there is no enlarging mass

Consider injury, acute mastitis, or breast abscess

Treat the cause (➔ p. 667)

Try self-help measures ± drug treatment (➔ p. 667). Refer if not settling

Urgent referral if unilateral and ≥50y
Consider discussion with a specialist if unilateral and <50y or bilateral discharge

Refer urgently if ≥50y
Consider discussion with a specialist if <50y and of new onset

Eczema—refer urgently if does not respond to topical treatment
Peau d'orange—refer urgently
Erythema—consider cellulitis, infected skin lesions, e.g. sebaceous cyst, mastitis, or breast abscess

Figure 19.1 Algorithm for management of breast symptoms

Management

- **No lump** Reassure. Educate the woman about breast awareness (➔ p. 662). Consider reviewing in 6wk
- **Discrete lump** Refer (urgently if aged ≥30y)
- **Asymmetrical nodularity**
  - ≥50y—consider discussion with a specialist/referral
  - <50y—review in 6wk. If the nodularity has gone, reassure; otherwise refer. If FH, have a low threshold for discussion with a specialist and/or referral

❗ Any patient being referred with a breast lump will be concerned about the possibility of breast cancer even though most will not have cancer.

Discharge from the nipple 90% of premenopausal women can express milky, multiple duct discharge. Ask about colour, quantity, and whether the discharge is unilateral/bilateral. Examine to check for lumps. Note colour/quantity of discharge and whether the discharge is coming from multiple or a single duct and is spontaneous or expressed.

Differential diagnosis

- Physiological (e.g. pregnancy)
- Duct ectasia—➔ p. 669
- Breast cancer—➔ p. 674
- Intraductal papilloma—➔ p. 669

Management

- Refer urgently if unilateral and ≥50y
- If <50y and any features suggesting pathological cause (Table 19.1), consider discussion with a specialist and/or referral

Breast pain or mastalgia Most common in women aged 30–50y. Use a pain chart for >2mo to distinguish cyclical from non-cyclical pain.

Cyclical breast pain Common. Two-thirds of women >35y have cyclical mastalgia which causes distress or interferes with lifestyle. Symptoms are often longstanding. *Features:*

- Usually bilateral though may not be the same intensity in both breasts
- Pain is generally felt over the lateral side of the breast, increases from mid-cycle onwards, and is relieved by menstruation

Examination may reveal tenderness ± areas of nodularity/lumpiness.

Table 19.1 Features of nipple discharge which suggest physiological or pathological cause

Physiological cause likely	Pathological cause more likely
Bilateral	Unilateral
Multiple ducts	Single duct
On expression only	Spontaneous
Green, milky	Red, brown, black
Stains only	Profuse and watery

Differential diagnosis

- Physiological
- Duct ectasia/periductal mastitis ➔ p. 669
- Breast cancer ➔ p. 674
- Sclerosing adenosis ➔ p. 668
- Mastitis ➔ p. 669 and p. 818
- Breast abscess ➔ p. 669
- Referred pain (e.g. cervical root pressure)

Management of mild/moderate cyclical pain 85% patients. Reassure that breast pain is a very *unusual* symptom of breast cancer. Explain the hormonal basis of symptoms. *Consider:*

- Diet Reducing saturated fats and caffeine may help
- Support Advise to wear a soft support bra at night
- OTC medication Try simple analgesia (e.g. paracetamol) and/or NSAID. Some women also find oil of evening primrose (gamolenic acid) effective, but it may take 4mo to work
- Changing/stopping hormonal contraceptives or HRT

Management of severe cyclical pain Defined as pain for >7d/mo for >6mo which interferes with lifestyle. Affects 15% patients. Try measures for management of mild/moderate cyclical pain first. If they fail, consider referral for specialist assessment.

Specialist treatments include: danazol, bromocriptine, tamoxifen, and LHRH analogues. Drug treatment helps ~80% of women. Treatment should be reviewed after 3–6mo and only continued if necessary. After stopping treatment, symptoms recur in about half, but are often less severe.

Non-cyclical breast pain Pain which is either continuous or intermittent but with no relationship to the menstrual cycle. Ask if the pain is localized or diffuse:

- Well-localized/point-specific pain Consider ill-fitting bras (especially underwired), breast cyst, breast abscess, mastitis, breast cancer (rarely presents with pain), chest wall causes, e.g. costochondritis
- More generalized pain Usually referred pain. Consider nerve root pain, post-herpetic neuralgia, lung disease

Management Treat the cause. If no cause can be found, refer for specialist assessment.

Eczema of the nipple Suspect underlying breast cancer. Refer urgently for specialist assessment if no response to topical treatment—➔ p. 664.

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral.
🖱 www.nice.org.uk/guidance/ng12

Benign breast disease

Most breast complaints are benign and have a physiological basis. Despite this, women with breast complaints tend to 'assume the worst' when a new problem is discovered. In most cases reassurance that there is nothing sinister underlying their symptoms is all that is required.

Mastalgia (breast pain) ➔ p. 666

Fibroadenoma An aberration of normal lobular development. Peak age 16–24y. 3 types: common, giant (>5cm diameter), and juvenile (adolescent girls). Present with a discrete, firm, non-tender, and highly mobile lump ('breast mouse'). Account for 13% of breast lumps.

Management Refer for confirmation of diagnosis—urgently if ≥30y (➔ p. 664). Diagnosis is confirmed with a combination of USS, mammography, and fine needle aspirate/core biopsy. If the lump is large (>4cm), the fibroadenoma is excised. In other groups, reassurance is usually all that is needed. 95% of fibroadenomas do not enlarge after diagnosis and 25% ↓ in size or disappear with time.

❗ Fibroadenomas may calcify in older women and give a characteristic appearance on mammograms.

Sclerosing adenosis Benign condition resulting from over-proliferation of the terminal duct lobules. It can cause recurring pain and/or result in a small, firm lump in the breast. Often detected incidentally on mammography as a calcified, 'stellate' abnormality. Always refer for confirmation of diagnosis—urgently if ≥30y. Treatment is symptomatic.

Phyllodes tumour Peak age 40–50y. 3 types—benign (most common), borderline malignant (uncommon), and malignant (rare).

Presentation and management Presents with a breast lump. Refer—urgently if ≥30y—for confirmation of diagnosis through a combination of USS, mammography, and fine needle aspirate/core biopsy. Treatment is always surgical with wide excision of the lump. Recurrence may occur.

Fat necrosis Usually history of injury ± bruising. As bruising settles, scarring results in a firm lump in the breast ± puckering of the skin. Most common in women with large breasts. Always refer—urgently if ≥30y—to a breast surgeon for triple assessment (USS, mammography, + fine needle aspiration/core biopsy). Once diagnosis is confirmed, no treatment is needed. The lump often disappears spontaneously.

Breast cyst Benign and fluid-filled. Cysts may be of any size, single or multiple. Most common >35y. Usually premenopausal women but may occur in postmenopausal women taking HRT. Presents as a firm, rounded lump which is not fixed and not associated with skin changes/skin tethering.

First breast cyst Refer for exclusion of malignancy—urgently if ≥30y. Diagnosis is confirmed with aspiration and/or USS and/or mammography.

Past history of breast cysts 30% of patients who have had a breast cyst develop another at a later date. If the lump is accessible, it is reasonable to attempt aspiration. There is no need to send aspirated fluid for cytology if the fluid is *not* bloodstained and lump completely resolves. Refer if the fluid aspirated is bloodstained; the lump does not disappear completely; the cyst refills; aspiration fails; or cytology reveals malignant or suspicious cells

⚠ Do not attempt aspiration if you have not been trained to do so as there is a small but significant risk of pneumothorax.

Galactocoele Milk-containing cyst which arises during pregnancy. Refer any new lump arising in pregnancy to a breast surgeon. Repeated aspiration may be needed. Resolves spontaneously.

Duct ectasia Occurs around the menopause. Ducts become blocked and secretions behind stagnate. Presents as discharge from ≥1 duct which may be bloodstained ± breast lump ± nipple retraction ('transverse slit' appearance) ± breast pain.

Management Refer for confirmation of diagnosis—urgently if ≥50y or lump and ≥30y. Usually no treatment is needed. Surgery may be required to confirm diagnosis, if discharge is troublesome, or to evert the nipple.

Periductal mastitis Infected subareolar ducts. Affects younger women than duct ectasia with peak age 32y. Presents with breast tenderness ± inflammation in the areolar area. May also have nipple discharge and/or retraction and/or an associated inflammatory mass/abscess.

Management Treat with antibiotics, e.g. flucloxacillin 500mg qds. Advise smokers that smoking can slow the healing process. If an abscess is present, refer for drainage. Refer if any residual inflammation or masses following treatment to exclude cancer. If recurrent infection, refer for consideration of surgery to remove the blocked duct

Mastitis in lactating women ➔ p. 818

Intraductal papilloma Benign, wart-like lump that forms within a duct just behind the areola. Perimenopausal women are more likely to have a single intraductal papilloma; younger women often have >1. May be bilateral. Presents with nipple discharge which may be bloodstained ± a subareolar lump/nodule (30%). Refer for confirmation of diagnosis—urgently if ≥30y and lump and/or ≥50y and nipple discharge. Usually excised.

Breast abscess Usually occurs in a lactating breast following mastitis; occasionally in a non-lactating breast in association with indrawn nipple, mammary duct ectasia, or local skin infection. Presents with gradual onset of pain in 1 breast segment with hot, tender swelling of the affected area.

Management Refer for surgical assessment. May be treated with repeated aspiration under ultrasound guidance or surgical incision and drainage.

Mammary duct fistula Fistula between a mammary duct and the skin. Usually a complication of a breast abscess. Refer for surgical excision.

Breast cancer screening

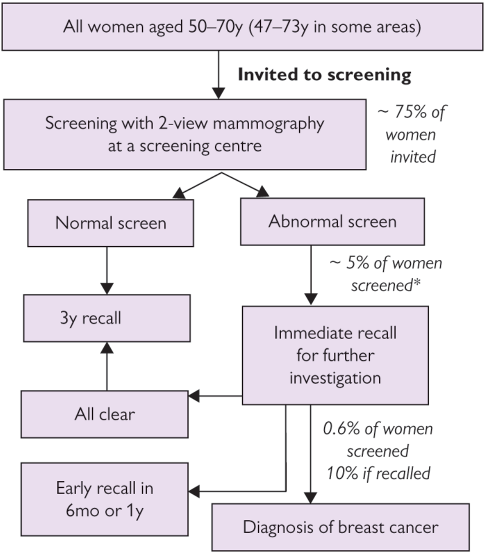
In the UK there has been a national screening programme for breast cancer since 1988. The aim of the programme is to detect breast cancer at an early stage in order to ↑ survival chances (stage I tumours—5y survival 84%; stage IV tumours—5y survival 18%).

Breast awareness Trials of self-examination have not ↓ mortality. Instead less formal 'Breast Awareness' is advocated—➔ p. 662.

Screening test

Low-risk women >47y 2-view mammographic screening is currently available to women aged 50–70y throughout the UK (in some areas age 47–73y as part of an evaluation trial). Older women can also request screening every 3y via their local breast screening unit. Screening detects 85% of cancers in women aged >50y (60% of which are impalpable) and ~70–80% screening-detected cancers have good prognosis. Organization of breast cancer screening in the UK—Figure 19.2.

High-risk women Women with family history of breast cancer may be at ↑ risk of breast cancer themselves (Figure 19.3, ➔ p. 673) and benefit from early/more frequent screening with either mammography or MRI—Table 19.2.


All women aged 50–70y (47–73y in some areas)
Invited to screening
Screening with 2-view mammography at a screening centre
~ 75% of women invited
Normal screen
Abnormal screen
~ 5% of women screened*
3y recall
Immediate recall for further investigation
All clear
0.6% of women screened
10% if recalled
Early recall in 6mo or 1y
Diagnosis of breast cancer

* 8% first time screens;
4% subsequent screens

Figure 19.2 Organization of breast cancer screening in the UK

Table 19.2 UK screening strategy for women at high risk of breast cancer

Risk group	Screening test		
	3-yearly mammography	Annual mammography	Annual MRI
TP53 mutation			20–49y; consider if 50–69y
BRCA1/2 mutation	≥70y	40–69y if not having MRI screening	30–49y
High risk—lifetime risk of >30%	≥60y	40–59y if not having MRI screening; consider if 30–39y and not having other screening	20–49y if >30% chance of *TP53* mutation; 30–49y if >30% chance of *BRCA1/2* mutation
Medium risk—lifetime risk of 17–30%	≥50y	40–49y; consider if 50–59y	

Interval cancers Cancer occurring in the interval between screens. Can occur through failure to detect a cancer at screening or as a result of a new event after screening took place. In the 1st year after screening, 20% of breast cancers are interval cancers. This ↑ to ~60% in the 3rd year.

Acceptability of screening See Table 19.3. ~81% women find mammography uncomfortable but 90% return for subsequent screens.

Table 19.3 Pros and cons of breast cancer screening

Benefits	Adverse effects
Earlier diagnosis Improved prognosis and lower mortality Less radical and invasive treatment needed Reassurance for those with −ve results	Discomfort and inconvenience of screening Radiation risks of screening False reassurance if false −ve results False reassurance if subsequent development of an interval cancer; possibly later presentation Anxiety and adverse effects of further investigation for those with false +ves Overdiagnosis of minor abnormalities that would never develop into breast cancer Earlier knowledge of disease and over-treatment for those for whom prognosis is unchanged

Further information

NHS Breast screening: programme overview. ✇ www.gov.uk/guidance/breast-screening-programme-overview
NICE (2013, updated 2017) Familial breast cancer. ✇ www.nice.org.uk/guidance/cg164

Risk factors for breast cancer

Breast cancer is now the most common cancer in the UK with ~55,200 new diagnoses every year (including ~390 new cases/y affecting men). Women have a 1 in 8 lifetime risk of developing breast cancer. Virtually all breast cancers are adenocarcinoma (85% ductal; 15% lobular).

Breast cancer screening ➔ p. 670

Risk factors

Geography More common in the developed world—migrants assume the risk of the host country within 2 generations.

Personal characteristics

- Age ↑ with age—~80% of breast cancers occur in women >50y
- Socioeconomic Higher incidence in more affluent social classes

Lifestyle factors

- Obesity ↑ risk post menopause
- Physical activity 30% ↓ risk if taking regular physical activity
- High-fat diet Probably associated with ↑ risk
- Alcohol ↑ risk by 7%/unit consumed/d

Reproductive history

- Early menarche or late menopause ↑ risk
- Pregnancy ↑ parity results in ↓ risk (32% ↓ risk in women reporting 3 births compared to women reporting 1); late age when first child is born and nulliparity ↑ risk
- Breastfeeding ↓ relative risk by 4.3% for each year of breastfeeding
- Combined hormonal contraception Slight ↑ risk (relative risk 1.24 for current users)—excess risk disappears within 10y of stopping
- Combined HRT 💣 Small ↑ risk if >51y and taking combined HRT (1 extra case/1000 women taking combined HRT/year) but no ↑ in breast cancer mortality. Excess risk disappears on stopping. Combined HRT ↑ breast density and the risk of having an abnormal mammogram. Oestrogen-only HRT does not ↑ breast cancer risk

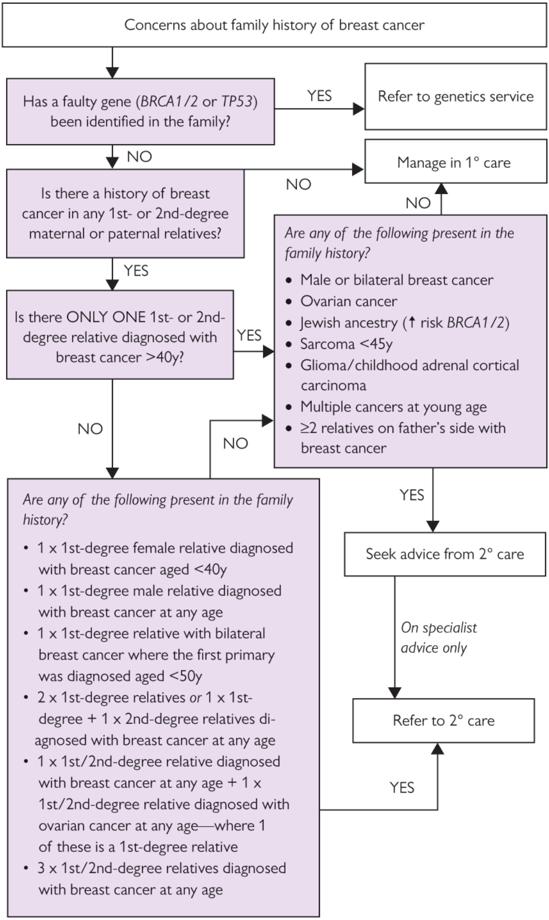
Other past medical history

- Past history of breast disease Ductal or lobular carcinoma *in situ*, florid hyperplasia, and papilloma with fibrovascular core all ↑ risk
- Ionizing radiation Exposure ↑ risk

Family history Referral algorithm for people with family but no personal history of breast cancer—Figure 19.3. Offer support (e.g. risk management advice, psychological counselling) for women with FH of breast cancer who are not eligible for referral and/or surveillance on the basis of age/risk level, but have ongoing concerns[N].

Further information

NICE (2013, updated 2017) Familial breast cancer. 🖱 www.nice.org.uk/guidance/cg164


Concerns about family history of breast cancer
Has a faulty gene (BRCA1/2 or TP53) been identified in the family?
YES
Refer to genetics service
NO
Manage in 1° care
Is there a history of breast cancer in any 1st- or 2nd-degree maternal or paternal relatives?
NO
NO
YES
Are any of the following present in the family history?
• Male or bilateral breast cancer
• Ovarian cancer
• Jewish ancestry (↑ risk BRCA1/2)
• Sarcoma <45y
• Glioma/childhood adrenal cortical carcinoma
• Multiple cancers at young age
• ≥2 relatives on father's side with breast cancer
Is there ONLY ONE 1st- or 2nd-degree relative diagnosed with breast cancer >40y?
YES
NO
NO
YES
Are any of the following present in the family history?
• 1 x 1st-degree female relative diagnosed with breast cancer aged <40y
• 1 x 1st-degree male relative diagnosed with breast cancer at any age
• 1 x 1st-degree relative with bilateral breast cancer where the first primary was diagnosed aged <50y
• 2 x 1st-degree relatives or 1 x 1st-degree + 1 x 2nd-degree relatives diagnosed with breast cancer at any age
• 1 x 1st/2nd-degree relative diagnosed with breast cancer at any age + 1 x 1st/2nd-degree relative diagnosed with ovarian cancer at any age—where 1 of these is a 1st-degree relative
• 3 x 1st/2nd-degree relatives diagnosed with breast cancer at any age
Seek advice from 2° care
On specialist advice only
Refer to 2° care
YES

❗ Family relationships:

- *1st-degree relative*—mother, father, sister, brother, daughter, son
- *2nd-degree relative*—grandparents, grandchildren, aunt, uncle, niece, nephew, half-sister, half-brother

Figure 19.3 Referral of women with a family history of breast cancer[N]

Breast cancer: diagnosis and care

Breast cancer screening ➔ p. 670

Prevention Consider referral to secondary/tertiary care if family history of breast cancer (➔ p. 673)

- Lifestyle measures—↓ alcohol intake; ↓ weight; ↑ exercise; avoid exogenous sex hormones (e.g. HRT); breastfeed
- Chemoprophylaxis—tamoxifen ↓ risk of breast cancer by 40% in high-risk women but use is limited by side effects (thromboembolism and endometrial carcinoma)—other drug trials are in progress
- Prophylactic surgery—↓ risk by 90% in very high-risk women

Presentation Most common site is upper-outer quadrant of the breast. Often found at breast screening (➔ p. 670). *Clinical presentations*:

- Breast lump (90%)
- Breast pain (21% present with painful lump; pain alone <1%)
- Nipple skin change (10%). Any red, scaly lesion or eczema around the nipple suggests *Paget's disease of the breast*—intraepidermal, intraductal cancer. Refer urgently if not clearing with topical treatment
- Family history (6%)
- Skin contour change (5%)
- Nipple discharge (3%)
- Rarely presents with distant metastases, e.g. bone pain
- In the elderly, may present with extensive local lesions

Management If suspected, refer for urgent assessment (<2wk) to a breast surgeon—see ➔ p. 664. Specialist investigation includes mammography, USS, ± fine needle aspiration or core biopsy. If diagnosis is confirmed, further investigations include tumour markers, and/or CT/MRI, liver USS and/or bone scan to evaluate spread.

Treatment Includes surgery (lumpectomy + sentinel node biopsy ± axillary clearance; mastectomy), endocrine therapy, radiotherapy, and/or chemotherapy.

Table 19.4 Classification of breast cancer stage

Stage	TNM equivalent	Features
In situ	Tis N0 M0	Non-invasive
I	T1 N0 M0	≤2cm diameter No LNs affected No spread beyond breast
II	T0–2 N1 M0 *or* T2/3 N0 M0	2–5cm diameter and/or LNs in axilla involved No evidence of spread beyond axilla
III	T0–2 N2 M0 *or* T3 N1/2 M0 *or* T4 any N M0 *or* Any T N3 M0 *or*	>5cm diameter LNs in axilla involved No evidence of spread beyond the axilla
IV	Any T/N M1	Any sized tumour LNs in axilla may be affected Distant metastases

Adjuvant endocrine therapy Oestrogen has an important role in the progression of breast cancer. Oestrogen and progesterone receptors determine the response to endocrine therapy.

- **Tamoxifen** ↑ survival of patients with oestrogen receptor +ve tumours (60% tumours) of any age but rarely causes endometrial cancer—warn patients to report any untoward vaginal bleeding. Continue tamoxifen for ≥5y—take advice from a specialist prior to stopping
- **Aromatase inhibitors** e.g. anastrozole, letrozole, and exemestane. Block synthesis of oestrogen. Superior efficacy when compared to tamoxifen for postmenopausal women with hormone-sensitive early breast cancer and first choice for postmenopausal women with advanced breast cancer. Continue for ≥5y—take advice from a specialist prior to stopping
- **Trastuzumab (Herceptin®)** Monoclonal antibody directed against HER2, a receptor found in 1 in 5 breast cancers. Affects division and growth of breast cancer cells. Treatment option for women with early HER2 +ve cancer at high risk of recurrence and women with advanced HER2 +ve breast cancer. Administered IV every 3wk for 1y

❗ Optimum treatment regimes for breast cancer change regularly and there are regional variations. Many women will be asked to participate in clinical trials to answer important questions about best treatments.

Prognosis 73% of women diagnosed with breast cancer now will live 10y; 64% live ≥20y.

- Recurrence is most likely <2y after treatment—late recurrences do occur but the longer since diagnosis, the less the chance of recurrence
- Prognosis for an individual depends on age (best prognosis if 50–69y), stage of disease (Table 19.4), grade of tumour, and oestrogen receptor status (oestrogen receptor −ve tumours have poorer prognosis)
- Women living in affluent areas have better survival rates than those in deprived areas

Psychological impact of breast cancer Depression, anxiety, marital problems, and sexual problems are common. Be sensitive. Discuss possibilities of reconstructive surgery or breast prostheses as appropriate. Refer to the specialist breast care nurse for support and advice.

Lymphoedema ➔ p. 1025

Further information

NICE (2009, updated 2017) Advanced breast cancer: diagnosis and treatment. 🖱 www.nice.org.uk/guidance/cg81

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

NICE (2018) Early and locally advanced breast cancer: diagnosis and treatment. 🖱 www.nice.org.uk/guidance/ng101

Information and support for patients

Breast Cancer Now ☎ 0333 20 70 300 🖱 http://breastcancernow.org/
Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk

Chapter 20

Gynaecology

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The menstrual cycle

A good working knowledge of the menstrual cycle is essential to understand its endocrine disorders and their management. One menstrual cycle lasts from the start of one period until the day before the start of the next. The average length of a cycle is 28d, but anything from 24 to 35d is common. The menstrual cycle is split into four (Figure 20.1).

Follicular or proliferative phase

Hormone changes Levels of oestrogen and progesterone are low. There is ↓ negative feedback on the pituitary as a result so follicle-stimulating hormone (FSH) levels ↑. FSH stimulates follicle development in the ovary. The developing follicles then produce oestrogen.

Changes within the reproductive organs

- Ovaries Follicles develop. One follicle becomes dominant
- Uterus Endometrium thickens (proliferates)
- Vagina Tends to be drier with thicker mucus

Ovulation Occurs halfway through a cycle (~14d before the next period). The dominant follicle ruptures and an egg is released into the fallopian tube. The follicle fills with blood after rupturing and there may be brief pain—'*mittelschmerz*' (reassure—no treatment needed). The egg travels along the fallopian tube into the uterus where it may be fertilized if the woman is sexually active and not using contraception.

Secretory or luteal phase

Hormone changes After ovulation, the ruptured follicle forms the corpus luteum (yellow body) and secretes progesterone and oestrogen.

Changes within the reproductive organs

- Ovaries Corpus luteum forms. If pregnancy does not occur, the corpus luteum begins to degenerate ~4d prior to menstruation
- Uterus Progesterone causes the endometrium to become ready to receive a fertilized egg. The endometrium becomes oedematous and more vascular; the glandular component becomes coiled/tortuous
- Vagina Mucus becomes thinner, more watery, and slippery. It becomes thicker again towards the next period as progesterone ↓
- Other changes Progesterone may cause 'water retention', breast tenderness, and mood changes

Periods (menstruation) With regression of the corpus luteum, oestrogen and progesterone levels ↓. This causes necrosis, bleeding, and sloughing of the endometrium → a period or menstruation. Periods begin aged 11–16y and continue until the menopause (usually 45–55y). Bleeding can last from 1 to 8d (average 5d) and is generally heaviest in the first 2d. Blood loss in each period is ~20–60mL (>80mL is abnormal and may lead to anaemia). Some period pain is common and normal.

Prolonged menstruation Bleeding for >5–6d/cycle. Most loss occurs in the first 3d. Long periods do not equate to ↑ menstrual loss, so prolonged menstruation per se does not need investigation. Frequently goes with menorrhagia—➜ p. 684.

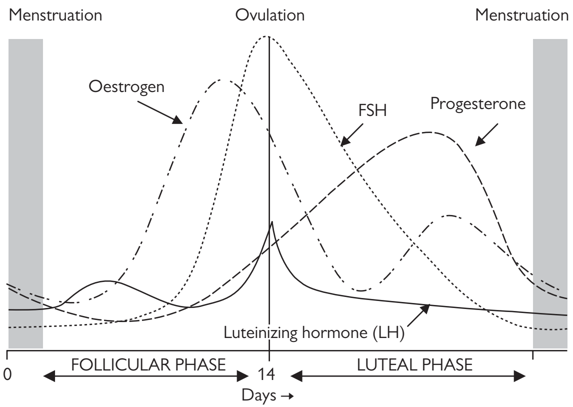

Figure 20.1 Hormone changes throughout the menstrual cycle

Anovulatory cycles Common around menarche, in the perimenopause, and in women with polycystic ovarian syndrome (PCOS). In the absence of ovulation, there is no luteal phase of the cycle leading to variable and erratic cycle length.

Postponing menstruation

- **Combined oral contraceptive (COC) pill** Started ≥1mo before and continued throughout the time the withdrawal bleed should have occurred (2 packets back-to-back without a break). The withdrawal bleed will occur after the second packet is finished
- **Combined contraceptive patch** Can be used for 6wk without a patch-free break to postpone a period
- **Oral progestogen** Norethisterone 5mg tds (licensed) or medroxyprogesterone 10–20mg/d (unlicensed) starting 3d before anticipated onset of menstruation. Menstruation occurs 2–3d after stopping

Odd colour/smell of menstrual blood No known associations.

Post-coital and intermenstrual bleeding

- **Post-coital bleeding (PCB)** Non-menstrual bleeding occurring during or after sexual intercourse. Consider cervical cancer (➔ p. 702). Other possible causes include infections of the lower genital tract, cervical ectropion and polyps, trauma, and vaginal and vulval lesions
- **Intermenstrual bleeding (IMB)** Vaginal bleeding at any time during the menstrual cycle other than menstruation. May be physiological or related to use of hormonal contraception. Other causes include: endometrial polyps, uterine fibroids, endometrial hyperplasia or cancer, endometritis, and cervical, vulval, or vaginal cancer

⚠ Always perform a full pelvic (including speculum) examination. If suggestive of cervical cancer, refer urgently (<2wk) for gynaecology opinion[N]. Do not use cervical smears as a diagnostic test. Consider urgent referral if intermenstrual bleeding even if the cervix looks normal.

Premenstrual syndrome

Most ♀ of reproductive age notice symptoms/bodily changes in the days/weeks leading up to their periods. These changes resolve or ↓ significantly during the period and are termed premenstrual tension (PMT), or premenstrual syndrome (PMS) if they occur on a regular basis and are severe enough to interfere with quality of life. >95% ♀ have some symptoms but <20% seek help. Debilitating symptoms occur in ~5%.

Cause Underlying mechanism is not fully understood; thought to relate to the hormonal changes that occur after ovulation affecting neurotransmitters in the brain.

Symptoms >100 symptoms described. The most common are:

- Psychological Mood swings, nervous tension, and/or irritability (when severe, termed *premenstrual dysphoric disorder*—PMDD)
- Physical Abdominal bloating, ↑ weight, breast tenderness, headache
- Behavioural ↓ visuospatial and cognitive ability, ↑ in accidents

Assessment^G Ask all women presenting with a history of PMT/PMS to keep a symptom diary for ≥2mo. Classify according to symptom pattern:

- Symptoms cyclical Relieved by menstruation and then recur after a symptom-free week—confirms PMS/PMT. If affecting quality of life, consider treatment. Otherwise, reassure and provide self-help information. If already taking medication containing progestogen (e.g. COC pill), consider changing the progestogen
- Symptoms cyclical but no symptom-free week Suggests premenstrual exacerbation of another underlying condition—treat the underlying condition ± PMS/PMT
- Non-cyclical symptoms Consider an alternative diagnosis

Lifestyle dietary modification

- Make allowances on days when symptoms are likely to be worst
- Wear loose clothes if feeling bloated
- Ensure adequate sleep and take regular exercise
- Eat regularly—some find small, frequent meals help; avoid sweet snacks between meals; make sure diet is low in fat/salt, caffeine, and alcohol, and contains plenty of fruit/vegetables and complex carbohydrate (e.g. bread, pasta, rice, potatoes)
- ↓ fluid intake or eat diuretic foods (e.g. strawberries, watermelon, aubergines, prunes, figs, parsley) to ease fluid retention
- OTC remedies may help (Table 20.1)

Primary care management^G

- Give lifestyle advice to all patients
- Consider further treatment if severe symptoms affecting quality of life and/or no response to diet/lifestyle measures
- Treatment—Table 20.1; base choice of treatment on symptoms
- For all treatments try a 3–6mo trial; ask women to keep a symptom diary. Be sure to follow up—as the first treatment may not work
- If symptoms are severe or primary care management is ineffective, refer to gynaecology/mental health services for specialist management

Secondary care treatment A multidisciplinary team approach is essential. Options include oestrogen patches + micronized progesterone; gonadotropin-releasing hormone (GnRH) analogues + HRT, or hysterectomy/oophorectomy + HRT.

Table 20.1 Treatment options for PMS^G

Treatment	Notes
Primary care treatment options	
Exercise	High-intensity exercise improves symptoms > low-intensity exercise
Cognitive behavioural therapy	Evidence that effective. Effects are smaller and slower than SSRIs—but more long-lasting
Vitamin B_6	Most studies suggest effective—advise women to take 10mg/d. High doses (>100mg/d) may cause reversible peripheral neuropathy
COC pill	Combined new-generation pills, given cyclically or continuously. COC containing drospirenone (e.g. Yasmin®) are recommended as first-line treatment
SSRIs (➔ p. 982)	Continuous or luteal phase (day 15–28) SSRI, e.g. sertraline—start at low dose and ↑ as needed. Warn about possible risks in pregnancy
Spironolactone	Effective for bloating/breast tenderness. Many women prefer 'natural' diuretics, e.g. Waterfall®, though there is no evidence of effectiveness
Complementary therapies 💣	Mixed evidence of effectiveness
Oil of evening primrose	May help breast tenderness. Can cause fits in patients with epilepsy
Chaste tree berry (*Vitex agnus-castus*)	Evidence of effectiveness is generally positive—can cause menstrual irregularity
Magnesium supplements	Evidence of effectiveness is generally positive. Used in the premenstrual phase
Calcium supplements	↓ symptoms including breast tenderness and swelling, headaches, migraine, and abdominal cramps
Relaxation/reflexology	Conflicting evidence—can do no harm

Further information

RCOG (2016) Management of premenstrual syndrome. 🖱 https://www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg48/

Information and support for patients and their partners

National Association for Premenstrual Syndrome ☎ 0844 815 7311 🖱 www.pms.org.uk

Amenorrhoea

Oligomenorrhoea Infrequent periods (>35d between periods). Manage as for amenorrhoea.

Primary amenorrhoea No menstruation by age 16y when growth and sexual development is normal. *Causes*:

- **Outflow abnormalities** Müllerian agenesis; transverse vaginal septum; androgen insensitivity (➔ p. 873), imperforate hymen, female genital mutilation
- **Ovarian disorders** Gonadal dysgenesis, e.g. Turner's syndrome—❗ gonads may have malignant potential
- **Pituitary disorders** Prolactinoma (➔ p. 340)
- **Hypothalamic disorders** Kallman's syndrome (congenital GnRH deficiency associated with anosmia)

Secondary amenorrhoea Absence of menses for ≥3mo in a previously menstruating woman. *Causes*: Figure 20.2.

History Always consider the possibility of pregnancy. *Symptoms*:

- Galactorrhoea—30% prolactinomas
- Weight change—weight ↓ or ↑ may cause amenorrhoea
- Hirsutism—may suggest PCOS or androgen-secreting tumour
- Life crisis or upset—e.g. exams, bereavement
- Exercise—high-intensity athletes (e.g. gymnasts)
- Sweats and/or flushes (suggests menopause)
- Cyclical pain—may suggest outflow obstruction
- Family history of premature menopause or late menarche
- Drug history—particularly contraceptives, e.g. injectable progestogens. Other drugs include: heroin, methadone, metoclopramide
- Past history of chemo- or radiotherapy or gynaecological surgery

Examination

- Weight and height—common if BMI $<19\text{kg/m}^2$
- External genitalia—structural abnormality, virilism
- Vaginal speculum examination—including cervical smear if overdue
- Pelvic examination—ovarian masses, uterine size
- General examination—2° sexual characteristics, hirsutism, systemic disease (including visual fields/retinal examination, ? prolactinoma)

❗ For young girls, replace pelvic/vaginal speculum examination with per-abdominal pelvic USS.

Investigation

- Blood— serum prolactin; TFTs; FSH/LH ; karyotype if phenotypical abnormality; serum testosterone if LH high, hirsutism, or virilism
- Transvaginal USS pelvis if structural abnormality or to confirm PCOS

Management of primary amenorrhoea Always refer for specialist assessment and treatment.

Management of secondary amenorrhoea

Physical exercise Explain reason for amenorrhoea—many women refuse to cut activity levels; consider HRT/CHC to protect bone density.

If underweight Investigate and treat reasons for weight loss (e.g. anorexia). Encourage weight ↑. If no response refer to gynaecology/eating disorders clinic.

Stress Reassure. Treat any psychiatric problems—periods should return spontaneously. Set a limit for return (e.g. another 3–4mo). If periods do not return, consider referral as there may be another cause.

Contraception Explain that some contraceptive methods do stop periods and reassure that this has no adverse effects:

- Injectable progestogens—periods usually return <1y after stopping
- Other hormonal methods—look for another cause of amenorrhoea if periods do not return ≤3mo after stopping

Endocrine

- Thyroid dysfunction—treat hyper- or hypothyroidism
- Hypothalamic causes—refer to endocrinology. After 6mo amenorrhoea, there is ↑ risk of coronary heart disease and osteoporosis. Consider use of HRT or CHC
- Hyperprolactinaemia—refer to endocrinology

Gynaecological

- Premature menopause—➔ p. 687
- PCOS—➔ p. 700

Cause not found Refer to gynaecology.

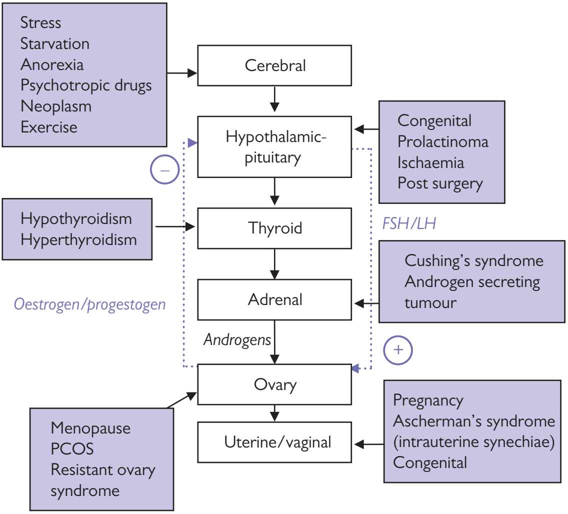
Stress
Starvation
Anorexia
Psychotropic drugs
Neoplasm
Exercise
Cerebral
Hypothalamic-pituitary
Congenital
Prolactinoma
Ischaemia
Post surgery
−
Hypothyroidism
Hyperthyroidism
Thyroid
FSH/LH
Cushing's syndrome
Androgen secreting tumour
Oestrogen/progestogen
Adrenal
Androgens
+
Ovary
Pregnancy
Ascherman's syndrome (intrauterine synechiae)
Congenital
Menopause
PCOS
Resistant ovary syndrome
Uterine/vaginal

Figure 20.2 Causes of amenorrhoea

Menorrhagia

Menorrhagia (heavy periods) is defined as menstrual loss ≥80mL/mo. 10% meet this criterion but 1 in 3 feel their loss is excessive. To gauge bleeding, ask about: number of tampons or pads used/d; use of double protection to prevent leaks; flooding and clots. Ask how periods affect life and activities. A menstrual diary may help. *Assessment*: Figure 20.3.

Differential diagnosis Physiological bleeding or dysfunctional uterine bleeding (50%). Exclude pregnancy. *Other causes*:

- Fibroids
- Congenital uterine abnormality, e.g. bicornuate uterus
- Pelvic infection
- Endometriosis
- Endometrial/cervical polyps
- Presence of IUCD
- Endometrial carcinoma
- Bleeding tendency/anticoagulant
- Hormone-producing tumours

❗ Hyper- or hypothyroidism, DM, prolactin disorders, adrenal, kidney, or liver disease, and some drugs can also cause menstrual disturbance.

Dysfunctional uterine bleeding (DUB) Excessive menstrual loss in the absence of any detectable abnormality.

Investigations Check FBC routinely. *Do not check*:

- Ferritin unless evidence of anaemia
- For coagulation disorders (e.g. von Willebrand's disease) unless heavy menstrual bleeding since periods started or personal/family history suggesting coagulation disorder
- Female hormone testing (LH/FSH) unless symptoms of menopause
- TFTs unless other symptoms/signs of thyroid disease

Primary care treatments Provide information about all available treatment options. Give iron supplements if anaemic:

Levonorgestrel-releasing IUS First-line treatment if:

- No identified pathology, *or*
- Fibroids <3cm diameter that are not distorting the uterine cavity, *or*
- Adenomyosis

Explain anticipated changes to bleeding pattern (irregular bleeding) can last up to 6mo. Wait >6mo to assess effects of treatment.

Other treatments Reassess after 2–3mo.

- **Non-hormonal** Tranexamic acid (1–1.5g tds) or NSAIDs (e.g. mefenamic acid 500mg tds). Start on day 1 and continue for days of heavy flow
- **Hormonal** CHC (➔ p. 730) or cyclical progestogen (e.g. norethisterone acetate 5mg tds from days 5–26 or medroxyprogesterone (MPA) 2.5–10mg/d for 5–10d beginning on days 16–21 of the cycle)

⚠ Management of very heavy bleeding

- Resuscitate as necessary—admit if shocked
- Reduce/stop bleeding with progestogen, e.g. norethisterone 5mg tds or MPA 10mg tds for 10d. Effective in 24–48h. A lighter bleed follows on stopping. An alternative is tranexamic acid 1–1.5g tds for 4d
- Correct anaemia and refer for specialist gynaecology assessment

Refer to gynaecology if

- Out-patient hysteroscopy is indicated (Figure 20.3)
- Fibroids >3cm diameter or distorting the uterine cavity
- Submucus fibroid—to consider hysteroscopic removal
- Other abnormality on USS that requires referral
- Primary care treatment has failed

❗ Offer NSAID/tranexamic acid while awaiting assessment/specialist treatment.

Secondary care treatment options include: endometrial ablation, embolization of fibroids, hysteroscopic resection of polyps/fibroids, myomectomy, or hysterectomy ± oophorectomy. GnRH analogues may be used prior to surgery.

Endometrial cancer ➔ p. 697

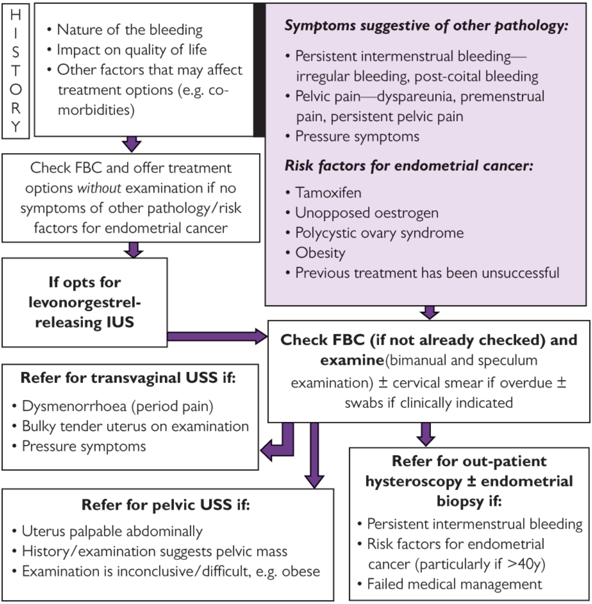
HISTORY
• Nature of the bleeding
• Impact on quality of life
• Other factors that may affect treatment options (e.g. co-morbidities)
Symptoms suggestive of other pathology:
• Persistent intermenstrual bleeding—irregular bleeding, post-coital bleeding
• Pelvic pain—dyspareunia, premenstrual pain, persistent pelvic pain
• Pressure symptoms
Risk factors for endometrial cancer:
• Tamoxifen
• Unopposed oestrogen
• Polycystic ovary syndrome
• Obesity
• Previous treatment has been unsuccessful
Check FBC and offer treatment options without examination if no symptoms of other pathology/risk factors for endometrial cancer
If opts for levonorgestrel-releasing IUS
Check FBC (if not already checked) and examine (bimanual and speculum examination) ± cervical smear if overdue ± swabs if clinically indicated
Refer for transvaginal USS if:
• Dysmenorrhoea (period pain)
• Bulky tender uterus on examination
• Pressure symptoms
Refer for pelvic USS if:
• Uterus palpable abdominally
• History/examination suggests pelvic mass
• Examination is inconclusive/difficult, e.g. obese
Refer for out-patient hysteroscopy ± endometrial biopsy if:
• Persistent intermenstrual bleeding
• Risk factors for endometrial cancer (particularly if >40y)
• Failed medical management

Figure 20.3 Assessment of patients with menorrhagia[N]

Further information

NICE (2018) Heavy menstrual bleeding 🖱 www.nice.org.uk/guidance/ng88

The menopause

From the Greek *meno* (month) and *pausis* (halt), menopause occurs when menstruation stops. Average age in the UK is ~51y (~2y sooner if smoker). Impact varies and depends on cultural, health, and social factors. Provide information about symptoms/management. Review 3mo after starting any treatment and then annually.

Diagnosis[N] For healthy ♀ >45y, diagnose without laboratory testing:

- **Perimenopause** If vasomotor symptoms and infrequent periods
- **Menopause** If no period for ≥12mo and not using hormonal contraception, or based on symptoms in ♀ without a uterus

Checking FSH FSH >30IU/L on 2 occasions >1mo apart suggests a ♀ is postmenopausal. *Check FSH if*: age <45y and menopause symptoms ± change in menstrual cycle. ❗ If having bleeds, check FSH from days 2–5 of the cycle; FSH levels may be normal in the perimenopause.

Could symptoms be due to another cause? Consider:

- Physical illness, e.g. thyroid disease, anaemia, DM, chronic renal disease
- Side effects of medication, e.g. Ca^{2+} antagonists cause flushing
- Social/psychiatric problems

Symptoms and management

Period changes

- **Changes in menstrual pattern** Common in the years before the menopause—typically cycle shortens after 40y by up to 7–10d. Cycle then lengthens—periods may occur at 2–3mo intervals until stopping
- **Dysfunctional uterine bleeding** Common in the perimenopause. Investigate if heavy, painful, irregular, intermenstrual, or post-coital bleeding
- **Late menstruation (>54y)** Investigate as ↑ risk of malignancy

Vasomotor symptoms 80% have hot flushes/sweats during the menopause—20% seek help. Often associated with palpitations. *Consider*:

- **Lifestyle changes and complementary therapies** Box 20.1
- **HRT** (➔ p. 688) First-line—effective for 80–90% of ♀
- **Non-hormonal treatments** SSRIs/SNRIs (e.g. sertraline 50mg od—unlicensed—useful if low mood); gabapentin (100–300mg tds—unlicensed); clonidine (50–75 micrograms bd—may ↓ BP). ❗ Do not offer fluoxetine or paroxetine to ♀ taking tamoxifen

Sexual dysfunction

- **Vaginal dryness and atrophy** Common. Manage with vaginal lubricants/moisturizers and/or topical oestrogen (even if already taking systemic HRT). Topical oestrogen at standard dose is thought safe even if history of breast cancer—check with supervising specialist if current/recent breast cancer; can be used continuously or intermittently
- **Loss of libido** Responds to administration of testosterone with HRT (unlicensed—50mg lasts 7–14d; apply daily to lower abdomen/upper thigh and rotate sites)

Urinary problems Common—incontinence, nocturia, and urgency. Stress incontinence does not respond to HRT but topical oestrogen may improve outcome of surgery. Recurrent UTIs and urge incontinence in older women ↓ with use of topical vaginal oestrogen.

Box 20.1 Lifestyle changes and complementary therapies

Lifestyle changes

- Low-intensity exercise (e.g. yoga)
- Avoiding trigger foods/drinks (e.g. spicy foods, caffeine, alcohol)
- Cool ambient temperature
- Wearing natural fibres
- Stress reduction

Complementary therapies ❗ May have unforeseen side effects, vary in amount of active ingredient, quality, and purity, and interact with other prescribed medications.

- **Black cohosh** Eases hot flushes—long-term effects are unknown
- **Isoflavones** (phyto-oestrogens, e.g. soy) May be helpful for flushes
- **St John's wort** May ease flushes/mood symptoms but interactions with antidepressants, anticoagulants, anticonvulsants, and tamoxifen
- **Red clover** May help—studies have mixed results; avoid with warfarin

💣 *Low mood* Consider social, physical, and cultural factors before offering HRT. Regular sustained aerobic exercise (e.g. swimming, running) ↓ psychological symptoms and insomnia; CBT may also be helpful. ❗ Do not treat with SSRI/SNRI unless clinical depression.

Musculoskeletal problems

- **Joint/muscle pains** Treat as needed with paracetamol ± NSAIDs
- **Osteoporosis** Consider HRT (➜ p. 688) to prevent osteoporosis in premature menopause. In older ♀, HRT is not recommended unless other reasons for prescribing HRT. Osteoporosis treatment—➜ p. 484

Contraception and the menopause Advise ♀ who are not using a hormonal method of contraception to continue using contraception after their LMP for 1y if aged >50y or 2y if aged <50y. Where timing of menopause is unclear due to use of contraception/HRT, advise women to continue contraception until age 55y and then stop. ❗ HRT is not contraceptive unless IUS is being used as the progestogen component.

Premature menopause (*premature ovarian insufficiency*) Menopause in a ♀ aged <40y. In most cases no cause is found. May be surgical (can follow hysterectomy even if ovaries are conserved), follow radio- or chemotherapy, or be associated with infection (e.g. mumps, TB) or other endocrine disease. Associated with ↑ all-cause mortality and ↑ risk of osteoporosis and CVD. HRT is recommended until the average age of menopause, i.e. 51y. Refer for specialist advice if HRT is contraindicated.

Further information

British Menopause Society 🖱 www.thebms.org.uk
NICE (2015) Menopause: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng23

Information and support for patients

Daisy Network (premature menopause) 🖱 www.daisynetwork.org.uk
Menopause Matters 🖱 www.menopausematters.co.uk

Hormone replacement therapy

HRT is very effective for the short-term relief of symptoms related to peri-menopause/menopause (➔ p. 686). It is also effective for prevention of osteoporosis in women who have had an early menopause (➔ p. 687).

Contraindications Undiagnosed abnormal vaginal bleeding; current or PMH of breast/endometrial cancer (topical vaginal oestrogen can be used); active liver disease with abnormal LFTs; porphyria cutanea tarda.

Choice of preparation Start with a low dose and provide a 3mo supply. Tablets, patches, and gels are available:

- **For women without a uterus** Give oestrogen alone, unless past history of endometriosis (endometrial foci may remain despite hysterectomy, so consider addition of a progestogen)
- **For women with an intact uterus** Progestogen is needed for the last 12–14d of the cycle or IUS to prevent endometrial proliferation. Alternatively, use a continuous oestrogen/progestogen preparation (not in the perimenopause or <12mo after last menstrual period)

Tibolone Oestrogenic, progestogenic, and weak androgenic action. Use in the same way as continuous combined HRT.

Topical vaginal preparations Oestrogen pessaries, creams, or rings. For vaginal dryness/atrophic vaginitis. Licence limits use to 3–6mo if uterus is present although commonly used for longer/continuously.

Things to do before starting HRT Ask: why does the woman want to start HRT? What are her expectations of treatment? Has she had a hysterectomy? If not, ask about bleeding pattern. Investigate abnormal bleeding prior to starting HRT. *Other points to cover*:

- Risk factors for osteoporosis, DVT, and CVD; FH of breast cancer
- Contraceptive requirement—➔ p. 747
- Drug history—previous experience of HRT; levothyroxine (may need to ↑ dose of levothyroxine after starting HRT—check TFTs); steroids (HRT ↓ effectiveness of steroids); antiepileptics (may ↑ elimination of oestrogen)

Examination Check BP; weight; check smear is up to date; consider examination for prolapse/vaginal abnormalities if symptoms. Advise women to check breasts and attend for breast cancer screening as eligible (➔ p. 662).

Starting HRT Explain risks of HRT (Table 20.2). Carefully balance risks against benefits for each individual. Support with health promotion information (e.g. smoking cessation advice, information about breast cancer screening). Review after 3mo. Ask if there has been any unscheduled PV bleeding. Check BP and weight. ↑ dose if symptoms are not controlled. *Common side effects*:

- **Oestrogen related** Fluid retention, breast enlargement and tenderness, nausea, headaches
- **Progestogen related** Headache, ↑ weight, bloating, and depression (↓ by changing to a preparation with a less androgenic progestogen, e.g. dydrogesterone or medroxyprogesterone)

Bleeding May be erratic for the first 2–3mo in patients taking cyclical HRT but should occur after the progestogen supplement in subsequent cycles. Continuous combined preparations may cause spotting for ~3mo.

⚠ Advise ♀ to report promptly if unscheduled bleeding occurs/continues >3mo after starting HRT. Investigate to exclude endometrial abnormality. If taking continuous combined HRT, consider changing to cyclical HRT.

Once established on HRT Review every 12mo and if any problems. Check BP, weight, breasts, symptoms, and bleeding pattern. Reassess risks and benefits.

Stopping HRT Consider stopping HRT after ~5y (or aged 51–52y if later). Discuss risks and benefits of ongoing HRT. Withdrawal can cause distressing flushes/sweats; offer a choice of gradually reducing or immediately stopping treatment. *Reasons to stop HRT immediately*:

- Sudden, severe chest pain, breathlessness/haemoptysis, or stomach pain
- Unexplained swelling or severe pain in calf of one leg
- Serious neurological side effects, e.g. severe headache, first fit
- Hepatitis, jaundice, or liver enlargement
- BP >160mmHg systolic and/or >95mmHg diastolic
- Prolonged immobility after surgery or leg injury
- Contraindication becomes apparent

HRT and surgery Stop HRT 4–6wk prior to surgery; restart only after full mobilization. If non-elective surgery, prophylaxis with LMWH and compression hosiery is needed.

Table 20.2 Risks of HRT ❗ Detailed information is available in the *BNF*

Risk	Notes
Venous thrombo-embolism (VTE)	↑ risk with oral not transdermal HRT Consider transdermal preparation if ↑ risk VTE, e.g. BMI >30kg/m^2, PMH/FH DVT or PE, severe varicose veins, immobility Seek specialist advice if strong FH or hereditary thrombophilia
Cardio-vascular disease (CVD)	Oestrogen alone—no or ↓ risk of CHD Oestrogen + progestogen—little or no ↑ risk of CHD Oral (but not transdermal) oestrogen—small ↑ risk of stroke Tibolone ↑ stroke risk ×2.2 from first year of treatment
Type 2 DM	HRT has no effect on glycaemic control or complications
Breast cancer	Oestrogen alone—no ↑ risk of breast cancer Oestrogen + progestogen and tibolone—↑ risk of breast cancer, related to treatment duration; risk ↓ to baseline <5y after stopping Radiological detection of breast cancer can be more difficult as mammographic density ↑ with HRT use
Other cancers	Endometrial cancer risk is ↑ if oestrogen-only HRT is used for women with a uterus. Risk is ↓ with addition of progesterone Risk of ovarian cancer is slightly ↑ but returns to baseline on stopping Risk of bowel cancer is ↓ with HRT use

Further information

NICE (2015) Menopause: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng23

Pelvic pain

Pelvic pain may be acute or chronic (pain for ≥6mo). *Causes*: Table 20.3.

History Allow the woman to tell her story. Ask about:

- **Pain** Site; severity; onset (?pregnant); character/timing/pattern (e.g. relationship to menstrual cycle or sexual intercourse, exacerbating/relieving factors, effects of movement/posture); other associated features. Decide whether the cause is gynaecological—patients usually have dyspareunia and pain may be cyclical
- **Bowel/bladder/psychological symptoms**
- **Past history** Ectopic pregnancy, pelvic infection/surgery, other factors (be sensitive to possible history of sexual abuse or rape)

Examination Abdominal, pelvic, and vaginal examination—including rectal examination if indicated and cervical smear if overdue.

Investigation Consider:

- **Urine** Pregnancy test, M,C&S, dipstick for protein, RBCs, nitrites, and leucocyte esterase
- **Blood** FBC, CRP
- **Radiology** Transvaginal pelvic USS if gynaecological cause is suspected
- **Screening for STIs** Offer to all sexually active women with chronic pelvic pain

Management

- **Acute pelvic pain** Admit if severe or if ectopic pregnancy cannot be excluded. Otherwise provide analgesia and treat the cause
- **Chronic pelvic pain** Affects 1 in 6 women in the UK. Often >1 cause—aim to identify and address contributory factors

Table 20.3 Causes of pelvic pain

Gynaecological		Non-gynaecological	
Acute	Chronic	Acute	Chronic
Ectopic pregnancy Pelvic inflammatory disease Endometriosis Torsion of fibroid Dysmenorrhoea Ovarian cyst (torsion, bleeding, abscess, or rupture)	Endometriosis Adhesions Fibroids Prolapse Ovarian cyst Venous congestion Pelvic inflammatory disease	Appendicitis Colitis Diverticulitis Cystitis Renal stones Neurological Psychological[a]	Irritable bowel syndrome (50%) Interstitial cystitis Musculoskeletal Psychological[a] Bowel or bladder cancer Nerve entrapment

[a] Psychological pain may be a consequence of and perpetuate physical pain. Diagnosis is one of exclusion.

Dyspareunia Pain on intercourse. 10% women admit sexual intercourse usually causes discomfort. It may be *superficial* (felt around the introitus) or *deep* (felt deep inside). There is a psychological element in most cases (a vicious cycle of pain → fear of intercourse which exacerbates symptoms). Address both physical and psychological aspects.

Superficial dyspareunia Examine if possible but do not insist. Treat the cause. If no specific treatment, try lidocaine gel. *Causes*:

- Vulval Vulvitis—atrophic, infective (candida, HSV); dystrophy; neoplasm; lichen sclerosus; lichen planus; vulvodynia
- Vaginal Vaginismus; lack of lubrication; vaginitis—atrophic, infective; congenital—imperforate hymen, atresia; surgery, e.g. painful episiotomy scar; contracture—atrophy or after surgery/radiotherapy
- Urethral Urethritis; urethral caruncle; urethral diverticulum

Deep dyspareunia Causes: endometriosis; pelvic inflammatory disease; retroverted uterus; ovarian mass (rarely ovarian cancer); non-gynaecological causes. Examine, treat any cause found; else refer for further investigation. If no cause is found or cause is untreatable, pain can be ↓ by limiting penetration. Often becomes a chronic problem.

Mittelschmerz ➔ p. 678

Dysmenorrhoea (painful periods) >50% premenopausal ♀ have pelvic discomfort around their period; interferes with lifestyle in ~10%.

Primary dysmenorrhoea No underlying pelvic pathology. Starts 6–12mo after menarche when ovulatory cycles are established. Presents with lower abdominal cramps ± backache which occur in the first 1–2d of each period. May be associated GI disturbance, e.g. diarrhoea/vomiting. Young ♀ (<2y after onset of menarche) with no other symptoms do not need examination unless pathology is suspected. Perform a full abdominal and pelvic examination if older or atypical features. Treatment is with:

- NSAID (e.g. mefenamic acid 500mg tds, ibuprofen 200–400mg tds)—effective in 80–90%; start with bleeding and continue until pain abates
- CHC or desogestrel POP—effective in 80–90%

❗ 10–20% do not respond—consider a missed cause; consider referral.

Secondary dysmenorrhea Suggests underlying pathology. *Causes*:

- Endometriosis/adenomyosis
- Chronic pelvic infection
- Cu-IUD/IUS
- Cervical stenosis
- Pelvic/abdominal surgery
- Intrauterine adhesions (Asherman's syndrome)
- Psychosexual problems

Starts later than teenage years, or may present as a change in pattern, type, or intensity of usual pain. Pain can start just before the period and last throughout. Often associated with deep dyspareunia ± other symptoms, e.g. abnormal bleeding, vaginal discharge.

Assessment and management of 2° dysmenorrhoea Do an abdominal, vaginal speculum, and bimanual pelvic examination. Look for tethered/fixed uterus, uterine tenderness, masses, and thickening in the posterior fornix (associated with dyspareunia and endometriosis). Do a cervical smear if overdue and offer STI screen if sexually active; consider referral for pelvic USS. Treat any underlying cause; else refer for further investigation (e.g. laparoscopy, hysteroscopy).

Further information

RCOG (2012) The initial management of chronic pelvic pain. 🖱 www.rcog.org.uk/globalassets/documents/guidelines/gtg_41.pdf

Endometriosis and adenomyosis

Endometriosis[N] Presence of tissue histologically similar to endometrium outside the uterine cavity and myometrium. Most commonly found in the pelvis but can occur anywhere. Affects 10–15% of women presenting with gynaecological symptoms. Ovarian deposits may result in *chocolate cysts* or *endometriomas*.

Risk factors Heavy periods; frequent cycles. ❗ Oral contraceptives and pregnancy are protective.

Theories of pathogenesis

- **Reflux and implantation** Menstrual loss flows backwards through fallopian tubes into the pelvis where it implants into the peritoneum and continues to grow under the influence of oestrogen
- **Transformation/induction** Peritoneal tissue transforms into endometrium under the influence of ovarian steroids or as a result of factors released when menstrual loss refluxes into the peritoneum
- **Mechanical transplantation** Endometrium transplanted from one location to another (e.g. during surgery) will grow at that new site
- **Vascular ± lymphatic spread** Thought to explain distant deposits, e.g. lungs, brain

Presentation Ask the woman to keep a symptom diary. Suspect endometriosis if ≥1 of the following symptoms/signs ± infertility:

- Chronic pelvic pain
- Period pain (dysmenorrhoea) affecting activities/quality of life
- Deep pain during/after sexual intercourse
- Cyclical (period-related) GI symptoms (e.g. painful bowel movements) or urinary symptoms (e.g. blood in urine, pain passing urine)

Examination May be normal. Offer abdominal and pelvic examination looking for abdominal masses and pelvic signs, e.g. pelvic masses, ↓ organ mobility, tender nodularity in the posterior vaginal fornix, and visible vaginal endometriotic lesions.

Investigation Offer sexually active women a STI screen. Refer for pelvic USS (transvaginal or abdominal) even if examination is normal. ❗ Do not exclude the possibility of endometriosis if USS is normal; if clinical suspicion, refer to gynaecology for further assessment/investigation.

Primary care management Treat on first presentation:

- **Pain** Try paracetamol ± NSAID (e.g. ibuprofen 400mg tds prn). If ineffective, a trial of neuropathic painkillers is worthwhile (➔ p. 188)
- **IUS** ↓ endometriosis pain with symptom control maintained >3y; depo-medroxyprogesterone acetate injection is an alternative
- **Drug treatment** If a woman is not trying to conceive and there is no evidence of a pelvic mass, try a progestogen (e.g. norethisterone 5mg bd or tds for ≥4–6mo—starting on day 1 to 5 of the cycle—if spotting occurs ↑ dose to 20–25mg/d and stop once bleeding has ceased), or continuous CHC (3 or 4 packets without a break then 7d break)
- **If symptoms are not controlled** Refer

Psychological support Many women with endometriosis have pain for years. Often there is delay in diagnosis of the cause. Be sympathetic and supportive and use a cooperative strategy for management.

Refer to gynaecology if

- Diagnosis is unclear after primary care assessment or patient requests referral—specialist options include laparoscopy and/or MRI
- Symptoms are not controlled with primary care management
- Severe, persistent, or recurrent symptoms of endometriosis
- Pelvic signs of endometriosis

Specialist treatments

- **Medical options** Include gestrinone and GnRH agonists (e.g. goserelin) ± HRT (↓ side effects and bone demineralization). Side effects can be troublesome
- **Surgical options** Include laparoscopy or laparotomy with ablation of lesions and division of dense adhesions; tubal surgery; hysterectomy. Laparoscopic ablation of mild endometriosis may ↑ fertility

❗ For all forms of specialist treatment there is a 15–20% recurrence rate.

Management of infertility Refer early for specialist care—➔ p. 750.

- If tubal damage—options are reconstructive surgery or IVF
- If no tubal damage—laparoscopic ablation may improve fertility

Chocolate cyst/endometrioma ➔ p. 700

Adenomyosis Usually affects multiparous premenopausal women aged >35y. Caused by extension of endometrial tissue and stroma into the uterine myometrium. May coexist with endometriosis (15%) but a separate entity.

Presentation May be asymptomatic, or present with dysmenorrhoea (pain often peaks near the end of menstruation), dyspareunia, and menorrhagia. On examination, the uterus may be symmetrically enlarged and tender. Refer for pelvic USS (transvaginal or abdominal) if suspected (Figure 20.3, ➔ p. 685).

Management No treatment needed if asymptomatic. Refer for further investigation if symptoms. Treatment is usually as for endometriosis, with hysterectomy ± bilateral salpingo-oophorectomy as a last resort. Medical treatment with GnRH analogues is a short-term option if severe symptoms and while awaiting surgery, but symptoms return once withdrawn unless the woman has reached the menopause in the interim.

Further information

NICE (2017) Endometriosis: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng73
NICE (2018) Heavy menstrual bleeding. 🖱 www.nice.org.uk/guidance/ng88

Information and support for patients

Endometriosis UK ☎ 0808 808 2227 🖱 www.endo.org.uk
Pelvic Pain Support Network 🖱 www.pelvicpain.org.uk

Prolapse

Prolapse occurs when pelvic organs herniate into the vagina due to poor pelvic muscle tone and weakness of pelvic ligaments. Affects 12–30% of multiparous and 2% of nulliparous women. Good obstetric practice ↓ risk. Risk is ↑ by:

- Childbirth
- Menopause
- Coughing and straining
- Congenital connective tissue disorders

Terminology Named according to the organs involved:

- Cystocele Bladder bulges into the vagina
- Urethrocele Urethra bulges into the vagina
- Rectocele Rectum bulges into the vagina
- Enterocele Loops of intestine bulge into the vagina
- Uterine prolapse Uterus descends into the vagina

Uterine prolapse is further classified by degree. The most dependent portion of the prolapse is assessed while straining:

- 1st degree Cervix remains in the vagina
- 2nd degree Cervix protrudes from vagina on coughing/straining
- 3rd degree (procidentia) Uterus lies outside the vagina; may ulcerate

Presentation Dragging sensation, feeling of 'something coming down', or a 'lump'. Symptoms are worse when upright, i.e. while awake, and exacerbated by standing for a long time, coughing, or straining.

Associated symptoms Depending on structures involved—stress incontinence (➔ p. 424), difficulty defecating, recurrent cystitis, and/or frequency of micturition.

Examination/investigation Check abdominal examination to exclude pelvic masses. Then in left lateral position with Sims speculum, ask the patient to bear down and watch the vaginal walls. Exclude pelvic mass by bimanual examination. Dipstick urine ± send for M,C&S.

Management Choice of treatment depends on patient preference, general health, degree of prolapse, severity of symptoms, and wish to preserve fertility and sexual activity. Options include:

- Lifestyle measures Weight ↓; smoking cessation
- General measures Treatment of coexisting conditions exacerbating prolapse, e.g. chronic cough due to COPD or asthma, constipation, menopause/atrophic vaginitis
- Physiotherapy Pelvic floor exercises (➔ p. 821). Refer to specialist physiotherapy if simple, self-help techniques fail
- Ring pessary Useful for those too frail for surgery, women who have symptoms but do not want surgery or as a temporary measure while awaiting surgery. Change pessary every 3–6mo. *Shelf pessaries* may be useful for women who cannot retain a ring pessary—consider referral
- Surgery Refer if the woman is fit for surgery and symptoms are of sufficient severity to warrant operation and/or she has incontinence and/or recurrent UTI. Surgical options include repair operations (anterior or posterior colporrhaphy), colpo-vaginal suspension, and hysterectomy (vaginal or abdominal)

Fitting a ring pessary

- Measure the approximate size required manually—the distance between posterior fornix and pubic bone can be measured roughly against the index finger
- Soften the ring in hot water and lubricate it well
- Insert the ring into the posterior fornix and tuck it above pubic bone
- Change the pessary every 3–6mo. Inspect the vagina for damage (e.g. ulceration) before washing and re-inserting the ring or inserting a new ring

Potential problems

- **Discomfort** Ring may be too big (try smaller size) or atrophic vaginitis (try topical oestrogen)
- **Infection** Remove, clear infection, then try again
- **Ulceration** Remove, allow to heal, consider alternatives, or reinsert when fully healed
- **Expulsion** Ring may be too small, pelvic musculature inadequate, or retropubic rim unsuitable

Mesh/tape complications Polypropylene mesh/tape is medical-grade plastic used for treatment of urinary stress incontinence and prolapse. The mesh/tape acts as a scaffold and is incorporated into the body via fibrosis. Mesh/tape complications usually occur <1y after surgery becoming increasingly rare after 2y. Extrusion is the most common complication, affecting ~5% of ♀ who have a mesh/tape implant.

Presentation of mesh/tape complications

- **Symptoms** Irregular vaginal bleeding/discharge; pelvic pain/swelling; discomfort during intercourse; recurrent new bladder/bowel symptoms; pain/prickling feeling in the vagina exacerbated by exercise; searing buttock/leg pain
- **Signs** Tenderness on palpating the mesh/tape; tape/mesh exposure in the vagina; recurrence of prolapse; vaginal adhesions/scarring

Management If mesh/tape implant but asymptomatic, reassure. Refer women who have had a mesh/tape implant and have symptoms/signs of a mesh/tape complication for specialist review.

Further information

NHS England (2017) Mesh oversight group report. 🖱 www.england.nhs.uk/wp-content/uploads/2017/07/mesh-oversight-group-report.pdf
NHS England (2017) Mesh complications leaflet. 🖱 www.england.nhs.uk/wp-content/uploads/2017/07/mesh-complications.pdf

Patient information

Association for Pelvic Organ Prolapse Support (APOPS) 🖱 www.pelvicorganprolapsesupport.org
Pelvic Mesh Owners Guide 🖱 https://pelvicmeshownersguide.com

Uterine problems

⚠ **Postmenopausal bleeding (PMB)**[N] Unexplained vaginal bleeding >12mo after menstruation has stopped because of the menopause.

Causes Atrophic change (most common); endometrial hyperplasia; endometrial polyps; endometritis; endometrial malignancy; cervical malignancy; fibroids; uterine sarcoma; non-genital causes (bladder/bowel).

Management Refer to gynaecology to be seen in <2wk if aged ≥55y and PMB; consider referral if <55y. Assessment involves transvaginal USS to assess endometrial thickness ± hysteroscopy/endometrial sampling.

⚠ **Pelvic/abdominal mass**[N] Refer to be seen in <2wk if abdominal/pelvic mass on clinical examination that is not obviously uterine fibroids.

Congenital abnormalities of the female genital tract

- **Uterine retroversion** 20% have a retroverted, retroflexed uterus. May be difficult to palpate bimanually
- **Duplication** Of the cervix and/or uterus; vaginal septum; bicornuate uterus (of varying degrees). Usually found incidentally. Refer as needed
- **Imperforate hymen** May cause cryptomenorrhoea which presents as 1° amenorrhoea (➔ p. 682). Refer for surgical release if suspected
- **Ambiguous genitalia** ➔ p. 873
- **Cervical incompetence** ➔ p. 768

Fibroids (uterine leiomyoma) Benign tumours of the smooth muscle of the myometrium affecting 1 in 5 women. Often multiple. *Risk factors*: nulliparity; obesity; FH of fibroids; African origin. Oestrogen dependent so regress post menopause. Named by location:

- Pedunculated
- Intramural
- Cervical
- Subserosal (bulge into peritoneum)
- Submucosal (bulge into endometrium)
- Separate from the uterus, e.g. in the broad ligament

Presentation Usually asymptomatic. May cause:

- **Pelvic pressure/discomfort and/or backache**
- **Menorrhagia** Often submucous fibroids distorting endometrial cavity
- **Pain** Torsion (pedunculated fibroid); degeneration. ‘Red degeneration’ may occur in pregnancy (pain, fever, and local tenderness)
- **Urinary symptoms** May press on the bladder causing ↑ frequency, a feeling of incomplete emptying, or difficulty passing urine
- **Infertility** May act as a ‘natural IUCD’
- **Problems in pregnancy** May cause abnormal lie and ↑ risk of PPH
- **Bulky uterus** ± pelvic mass felt abdominally

Pelvic USS is diagnostic. Check FBC if menorrhagia. ❗ Calcified fibroids may be an incidental finding on X-ray.

Management If asymptomatic/mild symptoms—no treatment is needed. Otherwise, management depends on symptoms, number, and location of fibroids, fertility plans, and patient preferences. Consider CHC or IUS to ↓ menstrual loss (➔ p. 684). Refer to gynaecology if fibroid(s) >3cm diameter, submucous, or distorting the uterine cavity, or primary care management does not control symptoms.

Secondary care treatments

- **Medical** GnRH analogues and selective progesterone receptor modulators (e.g. ulipristal acetate) cause fibroid shrinkage
- **Surgical** Uterine artery embolization; myomectomy (removal of fibroids only); hysteroscopic resection; hysterectomy

Endometrial hyperplasia Oestrogen causes endometrial proliferation; progesterone causes endometrial maturation; shedding follows withdrawal of oestrogen and progesterone. If oestrogen is given alone, the endometrium proliferates uncontrolled → irregular/heavy bleeding, polyps, and ↑ risk of endometrial carcinoma. Caused by anovulatory cycles (e.g. PCOS) or administration of unopposed oestrogen.

Endometrial cancer In the UK, ~9000 women each year are diagnosed with endometrial cancer (5% of all ♀ cancers). It is predominantly a disease of postmenopausal women with 93% of cases diagnosed in women >50y (peak age 75–79y). Risk is ↓ with current or past use of CHC and/or progestogens. Factors which ↑ risk include age and:

- Obesity
- Nulliparity
- Late menopause
- PCOS
- DM
- Drugs—unopposed oestrogen, tamoxifen
- Granulosa cell ovarian tumour
- FH—of breast, ovary, or colon cancer
- Previous pelvic irradiation
- Hereditary non-polyposis colorectal cancer

Presentation 15% are asymptomatic. Most present with PMB (75–80%). Premenopausally tends to occur in overweight women and present with continual bleeding. Rarely detected on cervical smear.

⚠ *Management*[N] Refer urgently to gynaecology (to be seen in <2wk) if endometrial thickness in a postmenopausal ♀ is >4mm on USS done for other reasons or PMB and ≥55y; consider referral if <55y. Consider endometrial cancer as a diagnosis and refer for urgent USS if ≥55y and:

- Unexplained symptoms of vaginal discharge presenting for the first time, or with thrombocytosis, or haematuria, *or*
- Visible haematuria and low Hb, thrombocytosis, or ↑ blood glucose

Treatment TAH and BSO ± radiotherapy and/or chemotherapy. Survival depends on age of the patient and stage/grade of the tumour. Stage I disease—85% 5y-survival; stage IV—25% 5y-survival.

Endometritis Acute infection of the endometrium, usually after surgery (including IUCD insertion) or childbirth. Presents with fever, low abdominal pain, uterine tenderness, and/or purulent discharge (may be bloodstained). Take high vaginal/endocervical swabs for M,C&S (including chlamydia). Treat with antibiotics, e.g. doxycycline 100mg bd for 7d + metronidazole 400mg bd for 1wk.

Pyometra This is a complication (uterine cavity fills with pus)—suspect if endometritis fails to clear and refer to gynaecology urgently.

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Ovarian disease

⚠ **Ascites or pelvic/abdominal mass**[N] Refer to be seen in <2wk if ascites or abdominal/pelvic mass on clinical examination that is not obviously uterine fibroids.

⚠ **Check CA 125**[N] If new irritable bowel syndrome aged >50y or if a woman (especially if aged >50y) reports unexplained weight ↓, fatigue, change in bowel habit, or any of the following >12×/mo:

- Persistent abdominal distension ('bloating')
- Feeling full ('early satiety') and/or loss of appetite
- Pelvic/abdominal pain
- ↑ urinary frequency/urgency

If CA 125 is ≥35IU/mL Refer for urgent USS. If suspected ovarian cancer on USS refer urgently to gynaecology to be seen in <2wk.

If CA 125 <35IU/mL Consider investigating other causes of symptoms. If no other clinical cause is apparent, advise the woman to return if symptoms become more frequent and/or persistent.

Causes of ↑ CA 125 ↑ in 1% of healthy individuals. *Other causes*:

- Cancer—ovarian cancer (↑ in 85% with epithelial ovarian cancer); other intra-abdominal cancers
- Non-malignant conditions—e.g. endometriosis, PID, menstruation, DM, CCF, pleural effusion, liver disease, diverticulitis, appendicitis, ascites

Ovarian tumours May be solid or cystic. ❗ Early tumours are often asymptomatic and may be an incidental finding on abdominal/pelvic examination or USS done for another reason. In ♀ of reproductive age, >80% are benign. In postmenopausal ♀, the proportion of malignant tumours is ~50%. Classified according to tissue of origin:

- Tumours of surface epithelium—60%—see Table 20.4
- Germ cell tumours—15–25%
- Gonadal stromal tumours—5–10%
- Metastatic (from breast, stomach, colon, or genital tract)—5–10%

Simple, physiological, or functional cysts Common and often incidental finding on USS in premenopausal women. May cause pain due to tension within the cyst, rupture, torsion, or bleeding into the cyst.

- Follicular cyst An ovarian follicle fails to rupture. Unilocular and can reach a diameter of ~10cm. Usually regress during the subsequent cycle. More common in women using progesterone-only contraception
- Luteal cyst Forms if excessive bleeding into the corpus luteum. May be tender, cause abdominal pain, and delay the next period

Management if premenopausal[G]

- Check CA 125 and refer to gynaecology if multilocular/solid elements or >7cm diameter—urgently (to be seen in <2wk) if CA 125 ≥35IU/mL. Check AFP, hCG, and LDH if aged <40y and complex ovarian mass
- If simple cyst 5–7cm diameter, ensure annual USS follow-up; if <5cm diameter, no follow-up is needed

Management if postmenopausal[G] Refer to gynaecology—urgently (to be seen in <2wk) if CA 125 ≥35IU/mL or any sinister features on USS.

Table 20.4 Tumours of surface epithelium

Type of tumour	Subtype	10y survival
Serous Peak age 30–40y; 20–50% of ovarian tumours; 30% bilateral	**Benign serous cystadenoma** 60% of serous tumours; 25% of all benign ovarian tumours	100%
	Borderline serous cystadenoma 10% of serous tumours	90–95%
	Malignant serous cystadenocarcinoma 35–50% of serous tumours; bilateral in 40–60%; 40–50% of all ovarian cancers—85% have spread outside the ovaries at the time of diagnosis; >50% are >15cm diameter at diagnosis	15%
Mucinous Can be large; often multilocular and may contain viscid mucin—if burst, *pseudomyxoma peritonei* can result (mucin-secreting cells spread throughout the peritoneum)	**Benign mucinous cystadenoma** Peak incidence aged 30–50y; 80% of mucinous tumours; bilateral in 5–10%; 20–25% of all benign ovarian tumours	100%
	Borderline mucinous cystadenoma 10% of mucinous tumours; bilateral in 10%	90–95%
	Malignant mucinous cystadenocarcinoma Peak age 40–70y; 10% of mucinous tumours; bilateral in 15–30%; 5–10% of all primary ovarian cancers; average diameter at diagnosis ~16cm	34%
Endometrioid—peak age 50–60y; 30–50% bilateral; benign tumours are rare; malignant tumours account for 20–25% of all ovarian cancers; 30% coexist with endometrial cancer; 10% coexist with endometriosis		
Clear cell (mesonephroid)—5% bilateral; 5–10% of all ovarian cancers; 25% coexist with endometriosis; associated with hypercalcaemia		
Brenner (transitional cell)—rare—2–3% of all ovarian tumours; >90% are benign. If malignant, have poor prognosis; <5% are bilateral; associated with mucinous cystadenoma and cystic teratoma in 1 in 10 cases		
Undifferentiated carcinoma—no characteristic histological features (<10%)		

Ovarian cysts in children Unusual. Often incidental finding on USS done for another reason. Refer premenarchal girls with an ovarian cyst >2cm for further assessment.

Epithelial ovarian cancer (EOC) 90% of ovarian cancers. *Incidence*: >7000 cases/y in the UK (2.5% of all cancers). Ovarian cancer accounts for 6% of ♀ deaths. In the UK, ~80% of patients with ovarian cancer have had symptoms for <4wk before seeing their GP. *Risk factors*:

- Age Peak age 50–70y; 85% ovarian cancers occur in ♀ >50y
- Family history 10% of cancers occur in women with FH. 3–4× ↑ risk if one first-degree relative with ovarian cancer—but only 40% of familial cancer is explained by known gene mutations (e.g. *BRCA1/2*)
- Reproductive factors Greater exposure to ovulation ↑ risk (e.g. infertility, nulliparity, low parity)
- Lifestyle ↑ risk with obesity and smoking (mucinous tumours only)
- Other medical problems Breast cancer aged <40y (4× ↑ risk); cervical cancer (if radiotherapy); endometriosis; benign ovarian cysts
- Other factors Height >1.7m; long-term perineal use of talcum powder (💣); asbestos exposure

Protective factors Pregnancy—the more pregnancies, the lower the risk; COC pill—↓ risk by 60%—protective effect is maintained >20y after the COC pill has been discontinued; breastfeeding—may ↓ risk by 20%; tubal ligation—↓ risk by 30–70%; hysterectomy—↓ risk by 50%.

Prevention Ovarian cancer fulfils some criteria for population screening. Trials are ongoing. Meanwhile, high-risk ♀ (with ≥2 same-side family members with breast/ovarian cancer aged <50y) may be referred for specialist genetic advice, in addition to the UK Familial Ovarian Cancer Registry and counselling, regular testing, ± bilateral salpingo-oophorectomy.

Treatment Specialist management is with laparotomy ± adjuvant treatment with chemotherapy. *Prognosis*: stage I—92% 5y survival; stage IV (distant metastases—40% new diagnoses)—6% 5y survival.

Germ-cell tumours e.g. mature teratoma (ovarian dermoid cyst), immature teratoma, dysgerminoma, endodermal sinus tumour (yolk sac tumour), mixed germ-cell tumour. Peak incidence in early 20s. Most are unilateral. Associated with ↑ AFP and ↑ β-hCG (both are used as tumour markers). Prognosis is good with the majority cured.

Sex-cord stromal tumours e.g. thecomas, fibromas, Sertoli/Leydig cell or granulosa cell tumours. Usually present early with symptoms of hormone production, e.g. precocious puberty, PMB, or virilism. Granulosa cell tumours are linked with endometrial hyperplasia/carcinoma.

Endometrioma ('chocolate cyst') Cystic mass arising from ectopic endometrial tissue within the ovary as a result of endometriosis (➔ p. 692). The cyst contains brown, tar-like fluid hence the name 'chocolate cyst'. Often densely adherent to surrounding structures (e.g. peritoneum, fallopian tubes, bowel) and may be associated with pelvic pain, dysmenorrhoea, dyspareunia, and infertility. Refer to gynaecology if detected on pelvic USS. Treatment is usually with surgical excision if the cyst is ≥3cm in diameter.

Polycystic ovarian syndrome (PCOS) Up to 1 in 3 premenopausal women have polycystic ovaries on USS—1 in 3 of those women have PCOS. Cause is unknown—often there is a family history. Diagnosis requires presence of ≥2 of:

- **Oligomenorrhoea** And/or anovulation
- **Hyperandrogenism** Clinical and/or biochemical
- **Polycystic ovaries** ≥12 follicles (2–9mm diameter) in each ovary and/or ovarian volume $>10\text{cm}^3$

Symptoms and signs May be asymptomatic or have ≥1 of:

- Menstrual irregularity—oligomenorrhoea/amenorrhoea (affects 67%—more common if BMI $\geq 30\text{kg/m}^2$), dysfunctional uterine bleeding
- Anovulatory infertility
- Central obesity
- Acne
- Hirsutism
- Male-pattern baldness

Investigations

- Blood: ↑ testosterone (>2.5nmol/L—if >4.8nmol/L, exclude other causes of androgen hypersecretion, e.g. tumour, Cushing's syndrome), ↓ sex hormone binding globulin (SHBG)
- Transvaginal (or if unacceptable, transabdominal) USS ovaries

Complications

- Insulin resistance—2× ↑ risk of DM
- ↑ CVD risk—central body fat distribution, obesity, ↑ BP, ↑ triglycerides, ↓ HDL; 3× ↑ stroke/TIA risk
- ↑ endometrial cancer risk
- Obstructive sleep apnoea

Management

- Encourage weight ↓ and ↑ exercise as appropriate
- If oligomenorrhoeic, consider progestogens to induce a withdrawal bleed every 3mo to ↓ risk of endometrial hyperplasia
- Consider CHC if no contraindications to regulate menstruation; COC with anti-androgen (e.g. drospirenone) may ↓ acne/hirsutism
- Consider offering annual HbA1c/FBG check—particularly if obese (BMI >30kg/m^2), FH of DM, or aged >40y. Screen pregnant women with PCOS for gestational DM with a GTT at <20wk gestation (➔ p. 786)
- Clomifene can be used to induce ovulation (➔ p. 751)—refer to specialist infertility services
- Metformin (unlicensed) may be helpful for insulin sensitivity and menstrual disturbance. Also used to ↑ fertility if clomifene has failed
- Hirsutism—➔ p. 312

Ovarian hyperstimulation Iatrogenic condition resulting from overstimulation of the ovaries in the course of infertility treatment.

- **Mild** >10% patients receiving gonadotrophin therapy. Abdominal pain/swelling ± vomiting/diarrhoea. Manage with rest and simple analgesia, e.g. ibuprofen or paracetamol prn
- **Severe** 1% patients receiving gonadotrophin therapy. Abdominal pain/distension, vomiting/diarrhoea, ascites, pleural effusion, and/or venous thrombosis. Admit

Further information

NICE (2011) Ovarian cancer: recognition and initial management. 🖱 www.nice.org.uk/guidance/cg122
NICE (2015, updated 2017) Suspected cancer: recognition and referral 🖱 www.nice.org.uk/guidance/ng12
RCOG (2010, reviewed 2017) Ovarian cysts in postmenopausal women. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg34/
RCOG (2011) Management of suspected ovarian masses in premenopausal women. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg62/
RCOG (2014) Long-term consequences of polycystic ovary syndrome. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg33/

Further information and support for patients

Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk
Ovacome ☎ 0800 008 7054 🖱 www.ovacome.org.uk
Verity Support for women with PCOS 🖱 www.verity-pcos.org.uk

Conditions of the cervix

⚠ **Refer urgently to gynaecology**[N] To be seen in <2wk if:

- Ascites or abdominal/pelvic mass on clinical examination that is not obviously uterine fibroids
- Aged ≥55y and postmenopausal bleed; consider referral if <55y

⚠ **Refer for urgent USS**[N] If ≥55y and:

- Unexplained symptoms of vaginal discharge presenting for the first time, or with thrombocytosis, or haematuria, *or*
- Visible haematuria and low Hb, thrombocytosis, or ↑ blood glucose

⚠ **Perform a full pelvic examination** Including speculum examination of the cervix, for patients with:

- Postmenopausal bleeding if <55y
- Alterations in menstrual cycle
- Intermenstrual bleeding
- Post-coital bleeding
- Vaginal discharge

Cervical intraepithelial neoplasia (CIN) Pre-malignant change of the cervical epithelium. The majority of these changes are found in asymptomatic women <45y (peak incidence 25–29y). CIN is a histological diagnosis resulting from biopsy—usually following an abnormal smear.

- CIN 1 Nuclear atypia confined to basal third of the epithelium. ❗ CIN 1 may revert to normality
- CIN 2 Nuclear atypia in basal two-thirds of the epithelium
- CIN 3 Severe dysplasia/carcinoma *in situ*—full-thickness epithelial nuclear abnormalities

Treatment Depends on grade of histological abnormality. For women with high-grade lesions (CIN 2/3) treatment options include ablation, excision (large loop excision of the transformation zone (LLETZ), knife cone biopsy), or hysterectomy.

Cervical cancer In the UK, ~3000 women/y are diagnosed with cervical cancer (2% of ♀ cancers). Only affects women who have been sexually active; >99.7% are associated with high-risk human papillomavirus (HPV) infection—most commonly types 16/18, but 14 high-risk types have been identified. Two peaks of incidence—women in their late 30s and 70s/80s. Most (80%) are squamous cell cancer, the rest adenocarcinoma. Incidence is ↓ as a result of cervical cancer screening, changes in sexual practices, and more recently HPV vaccination.

Risk factors All relate to likelihood of HPV infection:

- Low social class
- Smoking
- Early age of first intercourse
- Early age of first pregnancy
- Multiple sexual partners
- History of dyskaryosis
- Immunosuppression, HIV
- Contraception (↓ with barrier methods; ↑ if >5y COC use)

Presentation May be found on routine cervical screening. Symptoms include post-coital, intermenstrual, postmenopausal bleeding, and/or offensive vaginal discharge. Speculum examination may reveal cervical ulceration/mass or a cervix that bleeds easily.

Management Refer urgently to gynaecology. Treatment is with surgery ± radiotherapy depending on the stage of the disease:

- **Stage 1** Microinvasive cancer (A) or cancer confined in the cervix (B). 65% women present at stage 1. 5y survival ~80–99%
- **Stage 2** Invasion into the upper third of the vagina (A) or parametria (B) but not to the pelvic side wall. 5y survival ~60–90%
- **Stage 3** Extension to the lower third of the vagina (A) or pelvic side wall (B). 5y survival ~30–50%
- **Stage 4** Tumour involving bladder/rectum (A) or extrapelvic spread (B). 5y survival ~20%

Prevention of cervical cancer In the UK, girls and boys aged 12–13y are routinely vaccinated to protect them against most of the strains of HPV that cause cervical cancer—➔ p. 725.

Cervical screening ➔ p. 704

Cervical erosion/ectropion Physiological. An erosion or ectropion is the area of columnar epithelium visible within the vagina when the squamo-columnar junction moves down the cervix at times of high oestrogen exposure (e.g. pregnancy, CHC, puberty). Only treat if:

- Abnormal cervical smear, *or*
- Symptoms are causing problems, e.g. post-coital or intermenstrual bleeding, or excess discharge—refer to gynaecology for cautery

Nabothian cysts Cervical mucus retention cysts. Usually asymptomatic—no treatment needed. Refer for cautery if troublesome discharge.

Cervicitis Presents with vaginal discharge, intermenstrual/post-coital bleeding, and/or pain. Speculum examination shows mucopurulent discharge and inflamed, friable cervix. *Causes*: chlamydial infection (50%), gonococcus, and HSV. Investigate with swabs for M,C&S/NAAT as appropriate, and treat the cause.

Cervical polyps Develop from the endocervix and protrude into the vagina through the external os. Usually asymptomatic although there may be ↑ vaginal discharge and the lowest part of the polyp may ulcerate and bleed causing intermenstrual, postmenopausal, and/or post-coital bleeding. The vast majority are benign.

Treatment Avulsion (send for histology). Cauterize base with silver nitrate stick if necessary. Frequently recur. If postmenopausal, intermenstrual, or post-coital bleeding, refer to gynaecology to exclude other pathology.

Cervical incompetence ➔ p. 768

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Information and support for patients

Cancer Research UK ☎ 0808 800 4040 🖱 www.cancerresearchuk.org/about-cancer

Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk

Cervical cancer screening

In the UK, women and trans men who have retained a cervix are screened for cervical cancer from age 25 to 64y with 'smear' tests. Screening prevents ~1000–4000 deaths/y from squamous cell cervical cancer.

Taking a smear Ensure adequate training. Courses are available—update skills every 3y. Cells are collected from the cervix with a brush; the head of the brush is either broken off or rinsed into a vial containing preservative fluid before going to the laboratory for examination. Give all women information about the test, condition being sought, possible results of screening, and their implications.

Timing Avoid menstruation if possible (note on the request form if unavoidable). Ideal time is mid cycle. Routine bimanual examination is unnecessary—only do if clinically indicated (e.g. painful/heavy periods).

Screening intervals in the UK

- 3-yearly from 25 to 49y
- 5-yearly from 50 to 64y
- Annually—women with HIV infection

❗ Women older than the upper age for screening can be screened if they have never been screened previously or are under recall for previous abnormal results. Screening intervals in the UK are currently under review and may be lengthened.

Organization of the cervical screening programme Practices undertaking cervical screening must:

- Provide information to eligible women to allow them to make an informed decision about taking part in the programme
- Perform the cervical screening test (and ensure staff are properly trained and equipped to perform the test)
- Arrange for women to be informed about the results of their tests
- Ensure that results are followed up appropriately
- Maintain records of tests carried out, results, and follow-up

Primary high-risk HPV testing >99.7% of cervical cancers contain HPV DNA and women with HPV infection are 70× more likely to develop high-grade cervical abnormalities. 14 high-risk types have been identified. By 2020, all cervical smears in the UK will be tested initially for high-risk HPV. Those testing negative will be returned to normal recall without cytological examination (Figure 20.4).

Liquid-based cytology (LBC) Smear samples testing positive for high-risk HPV are examined for cytological changes. If the cytology is abnormal, the woman is referred for colposcopy. If normal, the smear is repeated after 12mo (Figure 20.4).

HPV vaccination and cervical cancer screening Women will continue to be invited for cervical screening when they reach the age of 25y, regardless of whether they have received HPV vaccination.

Other possible abnormalities seen on cervical smear

- **Dyskaryotic glandular cells** Refer for colposcopy
- **Atrophy** Common in peri-/postmenopausal women. No action

- **Endometrial cells** May be normal if IUCD *in situ*, hormonal treatment, or first half of 28d cycle. Otherwise, discuss with laboratory. Refer if reported as abnormal
- **Inflammatory changes** Common finding. Take chlamydial, gonococcal, and high vaginal swabs. Treat as necessary
- **Trichomonas, candida, or changes associated with HSV infection** Treat trichomonas or candida. Discuss any new diagnosis of HSV with the patient
- **Actinomyces** Associated with IUCDs—➜ p. 742

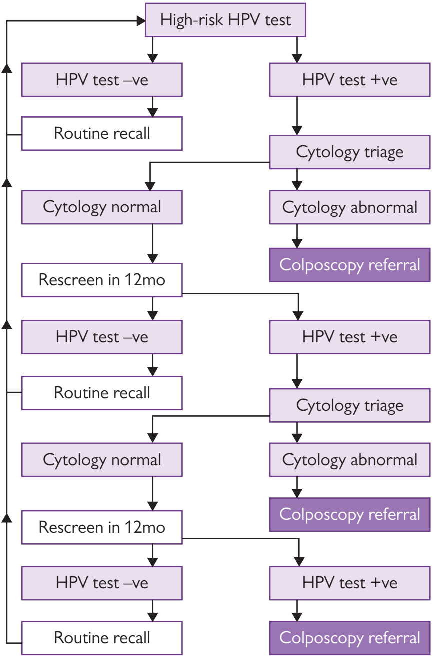
High-risk HPV test
HPV test –ve
HPV test +ve
Routine recall
Cytology triage
Cytology normal
Cytology abnormal
Colposcopy referral
Rescreen in 12mo
HPV test –ve
HPV test +ve
Routine recall
Cytology triage
Cytology normal
Cytology abnormal
Colposcopy referral
Rescreen in 12mo
HPV test –ve
HPV test +ve
Routine recall
Colposcopy referral

Figure 20.4 High-risk HPV primary screening for cervical cancer

Further information

Public Health England (2015) Cervical screening: programme overview. 🖱 www.gov.uk/guidance/cervical-screening-programme-overview

Information for patients

Public Health England (2018) Cervical screening: information leaflets. 🖱 www.gov.uk/government/collections/cervical-screening-information-leaflets

Vaginal and vulval problems

⚠ **Refer urgently to gynaecology**[N] (to be seen in <2wk) If:
- Unexplained vulval lump, ulceration, or bleeding
- Unexplained palpable mass in or at the entrance to the vagina

Symptoms/signs

- Vaginal discharge/infection—➔ p. 712
- Genital ulcers—➔ p. 711

Vulval itching (pruritus vulvae) ❗ Always exclude iron deficiency as a cause of itching. Treat the cause:
- Infection (e.g. candida, HSV, warts, threadworms, pubic lice, scabies)
- Skin conditions (e.g. lichen sclerosus, eczema, psoriasis)
- Vulval dystrophy/carcinoma
- Atrophic vulvitis
- Poor hygiene

Vulval lumps Common and usually benign.
- General causes Sebaceous cyst; varicose veins; haematoma; benign skin tumour (lipoma, papilloma, etc.); malignant skin tumour (1° or 2°)
- Specific causes Bartholin's gland cyst/abscess; urethral caruncle; endometriosis; carcinoma of the vulva; inguinal hernia

Atrophic vaginitis Vaginal soreness ± dyspareunia. Common cause of postmenopausal bleeding (PMB). Vagina looks pale and dry. Treat with topical oestrogen (e.g. each night for 2wk, then 2×/wk for ≥3mo) and/or consider systemic HRT. ❗ Refer women with PMB for specialist assessment—➔ p. 696.

Vaginal cysts May arise from remnants of the mesonephric ducts (anterolaterally) or following surgery or episiotomy (posterior, lower third). Usually no treatment is needed. If symptomatic or large, refer to gynaecology for assessment ± removal.

Benign vaginal tumours Benign leiomyomas or fibromyomas are uncommon. Refer for surgical removal.

Vaginal intraepithelial neoplasia (VAIN) Multifocal. Occurs in the upper third of the vagina—usually in association with CIN (➔ p. 702). May be asymptomatic or present with post-coital bleeding, PMB, or abnormal vaginal discharge. Treatment is by local ablation.

Vaginal cancer Rare—most common >70y; 90% are squamous cell—the rest clear cell (associated with *in utero* stilboestrol), secondary tumour, or sarcoma. Presents with PMB. Refer to gynaecology to be seen in <2wk. Treated with surgery (early stages) and/or radiotherapy.

Bartholin's gland swellings Painless vulval swelling due to obstruction of a Bartholin's gland duct → cyst formation. If infected, an abscess forms causing a painful, tender, red vulval lump. Cysts resolve spontaneously. Abscesses may resolve with antibiotics (if early) or discharge themselves. If not, admit for surgery (marsupialization).

Urethral caruncle Postmenopausal prolapse of the posterior urethral wall. Reddened area involving the posterior margin of the urethral opening. Usually asymptomatic—rarely bleeds or causes dyspareunia. Treat with topical oestrogen. Refer for surgery if symptoms persist.

Vulvodynia$^{\mathrm{G}}$ Chronic vulval pain without obvious cause. Common (4% of ♀) and may be generalized or localized. *Treatment*:

- **Topical** Avoid irritants, e.g. bubble bath—use soap substitute (e.g. aqueous cream) and topical moisturizer. Lidocaine gel/cream 5% may control pain during intercourse (apply 10min before)
- **Oral** Amitriptyline (10–75mg)—start with low dose and titrate ↑ slowly; if ineffective/side effects, consider gabapentin/pregabalin
- **Refer** If causing significant distress and not responding to treatment

Vulval dystrophy$^{\mathrm{G}}$ Associated with small risk of malignant change (<5%). Presents with vulval itching and/or soreness. Changes do not extend into the vagina. *Classification*:

- **Hypoplastic (lichen sclerosus)** Peak age 45–60y. Most common vulval dystrophy. 25% have FH/PMH of autoimmune disease, e.g. vitiligo, thyroid disease. Vulval skin looks atrophic ± white plaques (leukoplakia)
- **Hyperplastic** Usually affects postmenopausal women. There are multiple, symmetrical, thickened, hyperkeratotic lesions on the vulva

Management Map areas affected. Treat with topical steroid—e.g. clobetasol propionate ointment od for 1mo, then alternate days for 1mo, then 2×/wk for 1mo + barrier cream + soap substitute. Refer for biopsy if uncertain diagnosis, no improvement with treatment in <1mo or residual symptoms after 3mo, or other lesions develop (e.g. vulval lump, ulceration, or other skin changes). Symptoms may recur; re-treat as needed.

Vulval intraepithelial neoplasia (VIN)$^{\mathrm{G}}$ Some overlap with vulval dystrophy and may be associated with other genital tract neoplasia (e.g. CIN—➲ p. 702). Presents with abnormal vulval skin (pinky white + altered texture) ± white patches ± itch. Refer as for vulval dystrophy. Diagnosis is histological following skin biopsy and graded from VIN 1 (thickened epidermis—epithelial atypia in basal third only) to VIN 3 (carcinoma *in situ* 'Bowen's disease' with full-thickness nuclear atypia). Treatment depends on site, histology, and extent.

Vulval carcinoma$^{\mathrm{G}}$ Rare—most common aged >70y. Usually SCCs; others—melanoma, BCC, Bartholin's gland carcinoma, and adenocarcinoma. Generally occur on the labia and spread to local LNs. Present early with chronic pruritus vulvae, vulval lump, or ulcer. Refer for confirmation of diagnosis. Treatment is surgical—5y-survival rate ~95%.

Female genital mutilation ➲ p. 710

Further information

BAD (2018) Guidelines for the management of lichen sclerosus 🖱 www.bad.org.uk/shared/get-file.ashx?id=6039&itemtype=document
BASHH (2014) Vulval conditions 🖱 https://www.bashhguidelines.org/current-guidelines/skin-conditions/vulval-conditions-2014/
NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Information and support for patients

Association for Lichen Sclerosus and Vulval Health 🖱 www.lichensclerosus.org
Vulval Pain Society 🖱 www.vulvalpainsociety.org

Chapter 21

Sexual health and contraception

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Patient information about sexual health

NHS Guide to sexual health services 🖱 https://www.nhs.uk/using-the-nhs/nhs-services/sexual-health-services/guide-to-sexual-health-services/
Public Health England Sexwise 🖱 https://sexwise.fpa.org.uk/

Assessment of sexual health

Good communication skills are particularly important for clinicians when discussing sexual health problems and may improve health outcomes. Ensure a comfortable, private, and confidential environment.

Female genital mutilation (FGM) Common practice in many parts of the world. 4 types are specified by the World Health Organization:

- Type 1 Prepuce removal, or partial/total clitoris removal (clitoridectomy)
- Type 2 Removal of the clitoris + part/all of the labia minora (excision)
- Type 3 Removal of part/all of the labia minora with the labia majora either being sewn together covering the urethra and vagina leaving only a small opening for urine and menstrual fluid (infibulation)
- Type 4 Other: all other harmful procedures to the female genitalia for non-medical purposes, e.g. pricking, piercing, incising, scraping, and cauterizing

⚠ FGM is illegal in the UK. Healthcare professionals have a *mandatory* duty to report FGM in any ♀ aged <18y to the relevant authorities—➔ p. 902. If >18y, provide support and offer relevant follow up, if wanted. Where possible, identify other ♀ in the immediate/extended family (especially if <18y)—consider their risk of FGM and need for protection.

Sexual assault and rape ➔ p. 88

General assessment Objectives are to establish a constructive relationship with the patient to enable patient/doctor to communicate effectively and serve as the basis for any subsequent relationship to:

- Determine whether there is a sexual health problem and, if so, what
- Find out (where possible) what caused that problem
- Assess the patient's emotions and attitudes towards the problem. Be aware of signs of anxiety/distress. Recognize non-verbal cues
- Establish how it might be treated
- Assess contraceptive needs

History Use open questions at the start becoming directive when necessary—clarify, reflect, facilitate, listen. *Ask about*:

Presenting complaint Chronological account and concerns. If appropriate, ask about:

- Vaginal or urethral discharge
- Dysuria/other urinary symptoms
- Pain on intercourse (➔ p. 690)
- Erectile dysfunction (➔ p. 754)
- Genital/anal/perianal skin problems—soreness, itching, ulceration, warts
- Other symptoms, e.g. pelvic/abdominal/groin pain, postcoital bleeding, penile deformity, blood in the ejaculate, retrograde ejaculation

Past medical history

- Similar symptoms—for suspected sexually transmitted infections (STIs), ask about previous STIs, date of diagnosis, and treatment
- Obstetric history for women
- Urological problems and treatments or pelvic surgery
- Chronic medical problems—endocrine; cardiovascular; DM
- Medical treatment abroad—may be associated with ↑ HIV/hepatitis risk
- HIV testing/hepatitis B vaccination history

Drugs Prescription drugs (e.g. drugs associated with erectile dysfunction); illicit drugs (may be associated with erectile dysfunction, and history of injecting drug misuse is associated with ↑ hepatitis/HIV risk) and allergies.

Sexual history Current sexual partner (with whom had last sexual intercourse) and other recent sexual partners—if appropriate, ask about:

- Nature of relationship with partner—long-term partner; casual partner who could/could not be traced; paid-for partner; gender of partner
- Nature of intercourse—vaginal, anal, oral (?condom/dam use for oral sex)
- Contraception? Method used. Does the patient use a condom (male or female) regularly and consistently? Did it remain in place and intact?
- For women, establish date of LMP, cycle length, and regularity
- Symptoms in partner?

❗ For suspected STI, ask about all partners within the previous 3mo or incubation period of the suspected infection (if longer). If no partners are reported, note the last time the patient had sexual intercourse.

Social history

- Smoker?
- Alcohol consumption
- Use of drugs to ↓ sexual inhibitions ('*chemsex*')—associated with ↑ STI
- Travel abroad—if suspected STI, ask whether the patient had sexual intercourse abroad other than with their travelling partner, and with whom

Attitudes and beliefs

- How does the patient see the problem?
- What does he/she think is wrong?
- How does he/she think his/her partner views the situation?
- What does the patient want you to do about it?

Examination Examine the external genitalia and perianal area. Check groins for lymphadenopathy if STI is suspected. For ♀, perform pelvic and vaginal speculum examination. Consider digital rectal examination if indicated.

⚠ Explain the need for, and offer a suitably medically qualified chaperone for the examination of all patients. Record if a chaperone is declined.

Action Summarize the history back to the patient and give an opportunity for the patient to fill in any gaps. Check that the patient has no other concerns. Draw up a problem list and outline a management plan. Further investigations and interventions are guided by the findings on history and examination—so a good history and examination is essential. Then set a review date.

Genital ulcers *Causes*: genital herpes; primary syphilis; Behçet's syndrome. If history of foreign travel, partner from abroad, or doubt about diagnosis refer to sexual health clinic.

Further information

BASHH 2013 UK national guideline for consultations requiring sexual history taking. 🖱 www.bashhguidelines.org/media/1078/sexual-history-taking-guideline-2013-2.pdf

Royal College of Emergency Medicine (2017) A universal FGM flowchart and reporting tool. 🖱 www.rcem.ac.uk/docs/RCEM%20Guidance/FGM%20-%20BP%20Guide%20-%20Jul%202017.pdf

Vaginal discharge

All women have some vaginal discharge. Physiological discharge varies considerably and is affected by the menstrual cycle.

- **Before ovulation** Mucus is clearer, wetter, stretchy, and slippery
- **After ovulation** Mucus is thicker and stickier

Causes of 'abnormal' discharge 5 causes account for 95% cases:

- Excessive normal secretions
- Bacterial vaginosis (BV)
- *Candida albicans*
- Cervicitis (gonococcal, chlamydial, or herpetic)
- *Trichomonas vaginalis* (TV)

Rarer causes Cervical ectropion/polyp; Cu-IUD/IUS; chemical vaginitis (avoid perfumed or disinfectant bath additives and vaginal douches); foreign body (e.g. retained tampon—remove and treat with metronidazole 400mg tds for 7d); genital tract tumour; fistula.

History Ask about:

- **Symptoms** Vaginal discharge (itchy, offensive, colour, duration), vulval soreness and irritation, lower abdominal pain, dyspareunia, heavy periods, intermenstrual bleeding, fever, vulval pain
- **Sexual history** Recent sexual contact with new partner, multiple partners, presence of symptoms in partner, worries about STIs
- **Medical history** Pregnancy, diabetes mellitus, recent antibiotics
- **Attempts at self-medication**

Examination Always offer bimanual pelvic and speculum examination if:

- High risk for STI—age <25y; new sexual partner or >1 sexual partner in the past year; diagnosis of STI in the past 12mo
- Upper reproductive tract symptoms—abnormal bleeding (heavy, post-coital ± intermenstrual); pelvic/abdominal pain; deep dyspareunia; fever
- Pregnant, postpartum, or after miscarriage/termination
- After instrumentation (e.g. insertion of IUS/Cu-IUD, after colposcopy)
- Recurrent infection or failed treatment
- Requesting examination/STI testing

⚠ Do an abdominal, bimanual pelvic, and vaginal speculum examination. Look for tenderness on lower abdominal or bimanual palpation, cervical erosion/contact bleeding, discharge, foreign bodies, warts, or ulcers.

Investigation Consider checking pH of secretions with narrow-range pH paper. If >4.5, BV or TV is likely; pH is 4.5 with physiological discharge and candida infection. Other investigations to consider:

- High vaginal swab for M,C&S—only if symptoms/signs and/or pH consistent with specific diagnosis; the patient is pregnant, postpartum, or after miscarriage/termination; after instrumentation; or if there is recurrent infection/treatment has failed
- Vulvovaginal swabs for gonorrhoea and chlamydia
- Viral swab if herpes is suspected (if not available, refer to SH clinic)
- Opportunistic cervical smear if indicated (➔ p. 704)
- Self-taken vulvovaginal swab if examination is declined

Management Treat—if cause is unclear, refer to SH/gynaecology.

Sexually transmitted infections ➔ p. 714

Bacterial vaginosis (BV) Vaginal flora is changed from *Lactobacillus* species to anaerobes. Not sexually transmitted. Affects 10–40% of pre-menopausal women—about half are asymptomatic. *Associated with*:

- ↑ risk of preterm delivery (and ↓ risk if treated)
- Development of PID and endometritis following abortion or birth
- Infection after hysterectomy

Presentation Grey/white, thin, fishy-smelling, offensive discharge with no vulval soreness. On examination, the cervix looks normal; pH of secretions is >4.5. HVS for M,C&S may confirm diagnosis but treat without swab if no examination is carried out, or pH is >4.5 and typical clinical picture.

Management^{G} Without treatment, 50% remit spontaneously. Cure rate with all methods is ~85%. No need to treat the woman's partner. *Treatment*:

- Metronidazole 400mg bd for 5–7d or 2g single dose or clindamycin 2% cream 5g nocte PV for 1wk
- Recurrent infection—💣 suppressive therapy using metronidazole 400mg bd for 6d to cover the time of each period, metronidazole 0.75% gel 2×/wk PV for 4–6mo, lactic acid gel alternate nights for >1mo, and PV probiotics are all used but robust evidence of effectiveness is lacking

Candidiasis (thrush) Fungal infection—~20% of patients are asymptomatic. Predisposing factors include:

- Cushing's or Addison's disease
- Broad-spectrum antibiotics
- Radiotherapy/chemotherapy
- Immunosuppression
- Steroid treatment
- Vaginal trauma
- DM
- Pregnancy

Presentation Patient is well. Pruritus vulvae, superficial dyspareunia, and/or thick, creamy, non-offensive discharge. *Examination*: discharge (cottage cheese) and sore vulva which may be cracked/fissured. *Investigation*: usually unnecessary. If infection persists/recurs, send anterior fornix swab for M,C&S.

Management^{G} Only treat if symptomatic. Sexual transmission is minimal; there is no benefit from treating the partner unless overt infection:

- Try clotrimazole pessaries—cure rate ~90%
- Alternative is oral fluconazole 150mg stat, repeated after 3d if severe infection. Contraindicated in pregnancy or lactation—83% cure rate

Recurrent infection^{G} Advise loose, cotton underwear and avoidance of soaps, perfumes, or disinfectants in the bath. Consider vulval emollients to treat associated dermatitis. If 4 documented episodes (2 confirmed with microbiology) in a year, treat with fluconazole 150mg every 3d ×3, then 150mg weekly for 6mo.

Further information

BASHH (2007) Management of vulvovaginal candidiasis. 🖱 www.bashhguidelines.org/current-guidelines/vaginal-discharge/vulvovaginal-candidiasis-2007/

BASHH (2012) Management of bacterial vaginosis. 🖱 www.bashhguidelines.org/current-guidelines/vaginal-discharge/bacterial-vaginosis-2012/

RCGP (2013) Sexually transmitted infections in primary care. 🖱 www.rcgp.org.uk/-/media/Files/CIRC/RCGP-Sexually-Transmitted-Infections-in-Primary-Care-2013.ashx?la=en

Sexually transmitted infection

STI may present directly to GPs or be found incidentally (e.g. when doing a cervical smear). The easiest (and often best) option is to refer patients with suspected STI to a sexual health (SH) clinic.

Safe sex Advise to:

- Use condoms for all types of penetrative sex (vaginal, anal, oral sex)
- Have non-penetrative sex (e.g. body rubbing and mutual masturbation)
- Test for STIs before having sex with someone new, and suggest they are also tested
- Restrict number of partners to as few as possible
- Get vaccinated against certain infections e.g. hepatitis A/B (➔ p. 714) or take prophylactic antivirals to prevent HIV if in a high-risk group (➔ p. 720)
- Avoid sex when under the influence of alcohol/recreational drugs

Contact tracing Best done by SH clinics. If a patient refuses to go, provide a letter to give to contacts stating the disease involved, treatment given, and suggesting contacts visit their local SH clinic promptly.

High-risk groups[N] Include patients with STIs; men who have had sex with other men; people who have come from/visited areas of high HIV prevalence; substance/alcohol misuse; early onset of sexual activity; unprotected sex; and frequent change of/multiple partners. NICE recommends:

- Identification of high-risk patients opportunistically in general practice, e.g. at new patient checks, when attending for travel advice
- One-to-one discussions with those at ↑ risk of STIs lasting 15–20min, structured on the basis of behaviour change theories to ↓ risk-taking

Chlamydia screening ➔ p. 716 **HPV vaccination** ➔ p. 725

Prevention of HIV ➔ p. 720 **Vaginal discharge** ➔ p. 712

Acute pelvic inflammatory disease (PID) May be asymptomatic. Peak age 15–25y—especially if new partner and not using barrier contraception. Insertion of an IUD ↑ risk of PID for 21d after insertion. >10% develop tubal infertility after 1 episode; 50% after 3 episodes. Risk of ectopic pregnancy ↑ ×10 after a single episode. >50% of cases of PID are associated with STI—usually chlamydia (39%) or gonorrhoea (14%). Other causes include: *Gardnerella vaginalis*, anaerobes and *Mycoplasma genitalium*. Presents with:

- Fever and malaise
- Purulent vaginal discharge
- Acute pelvic pain (usually bilateral) and deep dyspareunia with bilateral lower abdominal tenderness, cervical excitation, and adnexal tenderness
- Abnormal vaginal bleeding—heavier periods, secondary dysmenorrhoea, intermenstrual and/or post-coital bleeding

Investigations Consider pregnancy test, swabs (HVS and endocervical swab for M,C&S and chlamydia/gonorrhoea screen), and blood tests (FBC—may show leucocytosis; ↑ ESR/CRP).

Management[G] Admit if unwell (fever >38°C, clinical signs of tubo-ovarian abscess or pelvic peritonitis), pregnant, or if ectopic pregnancy or other acute surgical emergency cannot be excluded. Otherwise advise rest and sexual abstinence; provide analgesia; treat with:

- Ofloxacin 400mg bd (or levofloxacin 500mg od) + metronidazole 400mg bd for 14d; or
- Ceftriaxone 500mg IM as a single dose, followed by oral doxycycline 100mg bd + oral metronidazole 400mg bd for 14d; or
- Oral moxifloxacin 400mg od for 14d (if *M. genitalium* isolated)

If the patient has an IUCD consider removal if symptoms are severe. If removed, advise about alternative contraception and emergency contraception if sexual intercourse <7d ago. If no improvement after 48h, admit; if slow recovery, consider referral for MRI/laparoscopy to exclude abscess formation.

Follow-up Arrange follow-up testing after 2–4wk to ensure clearance of infection. Arrange contact tracing via the SH clinic with treatment according to causative organism identified—if no causative organisms identified, offer ♂ partners treatment with doxycycline 100mg bd for 1wk.

Chronic PID Caused by inadequately treated acute PID. Presents with pelvic pain, dysmenorrhoea, dyspareunia (1 in 5) ± menorrhagia. *Examination*: lower abdominal/pelvic tenderness, cervical excitation ± adnexal mass. Screen for chlamydia and gonorrhoea. A −ve result does not exclude diagnosis. If chronic pelvic pain with no obvious cause, refer to gynaecology. Once diagnosis is confirmed, treatment options include long-term antibiotics or surgery.

Urethritis Inflammation of the ♂ urethra. Multifactorial condition primarily sexually acquired. Mucopurulent cervicitis is the ♀ equivalent. Characterized by discharge and/or dysuria although may be asymptomatic (found when a swab is taken following contact tracing). Treat the cause:

- **Gonococcal urethritis** *N. gonorrhoeae* is identified on urethral swab. Treat as for gonorrhoea (➔ p. 717)
- **Non-gonococcal urethritis**[G] Common organisms identified are: *Chlamydia* (11–50%); *Ureaplasma* (11–26%); *Mycoplasma genitalium* (6–50%); *Trichomonas vaginalis* (1–20%); adenoviruses (2–4%); HSV (2–3%). In 20–30% no organism is isolated. Treatment options are:

First-line		*If persistent/recurrent symptoms*
Doxycycline 100mg bd for 7d	→	Azithromycin 500mg stat then 250mg od for 4d + metronidazole 400mg bd for 5d
Azithromycin 500mg stat then 250mg od for 4d	→ ↘	Moxifloxacin 400mg od for 10–14d + metronidazole 400mg bd for 5d
Ofloxacin 200mg bd or 400mg od for 7d	→	Doxycycline 100mg bd for 7d +metronidazole 400mg bd for 5d

Further information

BASHH (2015, updated 2017) Management of non-gonococcal urethritis. 🖱 https://www.bashhguidelines.org/current-guidelines/urethritis-and-cervicitis/ngu-2015/

BASHH (2018) Management of pelvic inflammatory disease. 🖱 www.bashhguidelines.org/media/1170/pid-2018.pdf

BASHH/British HIV Association Safer sex advice. 🖱 www.bhiva.org/safer-sex-guidelines

NICE (2007) Sexually transmitted infections and under-18 conceptions: prevention. 🖱 www.nice.org.uk/guidance/ph3

Chlamydia, trichomonas, and gonorrhoea

Neonatal infection Babies may acquire sexually transmitted infections as they pass through the birth canal.

Ophthalmia neonatorum is conjunctivitis in an infant aged <30d. Most commonly caused by *Chlamydia trachomatis* or *Neisseria gonorrhoeae*. Send swabs for nucleic acid amplification testing (NAAT) and M,C&S. Seek immediate specialist advice if confirmed.

C. trachomatis Can also cause neonatal pneumonia (aged 1–3mo); otitis media; and pharyngitis—seek specialist advice if suspected.

Chlamydia Major cause of pelvic pain and infertility in ♀. *Presentation*:

- **Men** Usually asymptomatic. May have urethritis. Send urethral swab or first-catch urine sample for NAAT to confirm diagnosis. Lymphogranuloma venereum—➔ p. 722
- **Women** >70% are asymptomatic. Symptoms: vaginal discharge (30%); post-coital or intermenstrual bleeding; PID (10–30%—➔ p. 714); dysuria. Examination: mucopurulent cervicitis; hyperaemia and oedema of the cervix ± contact bleeding; tender adnexae; cervical excitation. Send vulvovaginal swab (can be self-taken) for NAAT to confirm diagnosis
- **Extra-genital infection** Rectal (asymptomatic in most; may cause anal discharge and/or rectal discomfort); pharyngeal (usually asymptomatic); or conjunctivitis (low-grade irritation). Send appropriate swabs for NAAT

❗ If exposed to potential infection <2wk previously, repeat test >2wk after exposure. A single individual may have more than one STI.

Management^{G} Contact tracing is essential to avoid reinfection; advise patients to avoid sexual intercourse until 7d after they and their partner(s) have started treatment. First-line treatment is with doxycycline 100mg bd for 7d. If allergy or pregnant/breastfeeding, azithromycin 1g stat followed by 500mg od for 2d is an alternative.

Follow up^{G} Carry out test of cure ≥3wk after treatment if pregnant or confirmed rectal infection. If aged <25y, or ≥25y and at high risk of reinfection, re-test 3–6mo after treatment.

Screening Chlamydia is a preventable cause of infertility, ectopic pregnancy, and pelvic inflammatory disease (PID). In the UK, there is a screening programme using urine or self-taken swabs, aimed at people aged <25y, to ↓ incidence/prevalence of PID. Self-test kits are available by post and through GP surgeries, sexual health/contraception clinics, and community venues (e.g. schools).

Trichomonas vaginalis (TV) *Presentation*:

- **Men** Asymptomatic (15–50%); urethral discharge; dysuria. Take urethral swab and first-void urine for M,C&S and NAAT
- **Women** 10–50% are asymptomatic. Symptoms: vaginal discharge, vulval itching, dysuria, offensive odour, low abdominal discomfort, vulval ulceration. Examination: 5–15% have no abnormalities; common findings include: vaginal discharge (~70%; classical frothy yellow discharge in 10–30%); vulvitis; vaginitis; 2% have classic 'strawberry cervix'. Send HVS from the posterior fornix at the time of speculum examination for M,C&S and NAAT. ❗ TV may be detected incidentally on cervical smear or self-taken swab

Management^G Refer to SH clinic for contact tracing. Advise to avoid sexual intercourse until patients and their partner(s) have completed treatment. First-line treatment is with metronidazole (400–500mg bd po for 5–7d or 2g po stat)—tinidazole 2g po stat is an alternative. If treatment fails, repeat 7d course of metronidazole. If second course fails, try metronidazole or tinidazole 2g po daily for 7–10d or metronidazole 800mg tds po for 7d.

❗ TV infection in pregnancy is associated with preterm delivery, low birth weight, and maternal postpartum sepsis but there is currently no evidence that screening and treatment of infected pregnant women ↓ this risk.

Gonorrhoea *Transmission*: ♂—1 in 5 exposures; ♀—1 in 2 exposures.

Presentation in men Depends on site of infection:

- Urethral infection >90% are symptomatic: urethral discharge (80%); dysuria (50%) 2–5d after exposure; prostatitis; urethral stricture
- Rectal infection Usually asymptomatic. Anal discharge (12%); anal/perianal pain or discomfort (7%)
- Pharyngeal infection >90% asymptomatic

If asymptomatic—send first-catch urine sample for NAAT; if symptomatic send urethral ± rectal ± pharyngeal swabs (as appropriate) for NAAT and M,C&S to confirm diagnosis.

Presentation in women Infection may be asymptomatic (50%) or cause vaginal discharge (50%), lower abdominal pain; dysuria but not frequency (25%); abnormal vaginal bleeding; PID; abscess of Bartholin's gland; miscarriage; or preterm labour. Rectal and pharyngeal infections are usually asymptomatic. If asymptomatic—send self-taken vulvovaginal swab for NAAT; if symptomatic—send endocervical swab ± rectal ± pharyngeal swab (as appropriate) for NAAT and M,C&S to confirm diagnosis.

Management^G Refer to SH clinic for contact tracing. Treat with ceftriaxone 500mg IM stat + azithromycin 1g po stat. If persisting symptoms/signs after treatment, test for cure with swabs sent for M,C&S >72h after completion of therapy. If asymptomatic following treatment, test for cure 2wk after treatment completion with NAAT, followed by culture if NAAT positive. If persistent infection, seek specialist advice.

Further information

BASHH 2015 UK national guideline for the management of infection with *Chlamydia trachomatis*. 🖱 www.bashhguidelines.org/media/1192/ct-2015.pdf

BASHH (2018) Update on the treatment of *Chlamydia trachomatis* (CT) infection. 🖱 www.bashhguidelines.org/media/1191/update-on-the-treatment-of-chlamydia-trachomatis-infection-final-16-9-18.pdf

BASHH United Kingdom national guideline on the management of *Trichomonas vaginalis* 2014. 🖱 www.bashhguidelines.org/media/1042/tv_2014-ijstda.pdf

BASHH UK national guideline for the management of gonorrhoea in adults, 2011. 🖱 www.bashhguidelines.org/media/1044/gc-2011.pdf

National Chlamydia Screening Programme 🖱 www.gov.uk/government/collections/national-chlamydia-screening-programme-ncsp

Hepatitis B and C

⚠ Hepatitis is a notifiable disease in the UK.

Hepatitis A (HBA) ➔ p. 394

Hepatitis B (HBV) Endemic in Asia. 3 major structural antigens: surface (HBsAg), core (HBcAg), and e antigen (HBeAg). Spread via infected blood, sexual intercourse, from mother → newborn, or human bites. High-risk groups—Box 21.1. *Incubation*: 6–23wk (average 17wk). Asymptomatic or fever, malaise, fatigue, arthralgia, urticaria, pale stools, dark urine, and/or jaundice. <1% develop acute liver failure; >85% recover fully; ~10% become carriers/have chronic hepatitis—which may cause cirrhosis ± liver cancer.

Investigation LFTs (hepatic jaundice—↑ bilirubin, ↑ ALT/AST, ↑ alkaline phosphatase), hepatitis serology (Figure 21.1).

- **HBsAg** Present from 1–6mo after exposure. Carrier if present >6mo
- **HBeAg** Present from 6wk–3mo after acute illness. Indicates high infectivity
- **Anti-HBs** Antibodies appear >10mo after infection; imply immunity

Management Advise about safe sex (until non-infectious or partner has been immunized and has sufficient immunity), to avoid alcohol and not to donate organs/blood/semen. Refer for specialist care. Treatment is supportive for acute illness ± antiviral agents if severe. Chronic hepatitis is treated with interferon, entecavir, or tenofovir (TDF or TAF).

Prevention Give hepatitis B immunoglobulin (HBIG) <2d after exposure to confer passive immunity to non-immune, high-risk contacts of infected patients (no effect after 7d). Hepatitis B vaccination is given as part of the routine UK childhood vaccination schedule for babies aged 2, 3, and 4mo. Babies born to hepatitis B +ve mothers are immunized <24h after birth (± HBIG). For high-risk groups (Box 21.1), use an accelerated regimen with vaccination at 0, 7, and 21d and a booster dose at 12mo (sooner if antibody response is <10IU/L after 4–6wk); patients with HIV infection may require an ↑ dose. Give hepatitis A vaccination if confirmed hepatitis B but not immune to hepatitis A.

Hepatitis C (HCV) Common. Spread is usually via contact with infected blood but can be mother → baby. Not easily spread through sexual contact. No source is found in 10%. *Incubation*: 2–25wk (mean 8wk). Acute infection is usually asymptomatic (>60%). Consider screening high-risk groups (Box 21.1). 50–85% develop chronic infection; without treatment, chronic infection → severe liver disease (30%) ± liver cancer (14%; 33% of those with cirrhosis).

Box 21.1 High-risk groups for hepatitis B and C infection

- Multiple sexual partners
- Men having sex with men (MSM)
- HIV +ve
- Intravenous drug users
- Prison inmates/workers
- Sex workers
- Healthcare workers
- Family contacts of a case/carrier
- Babies of hepatitis B/C +ve mothers
- Travellers to ↑ risk countries
- Adopted child from outside the UK
- Foster parents
- Blood transfusion before 1990
- People receiving regular blood products and their carers
- Patients on haemodialysis
- Staff/residents of residential homes for people with intellectual disability

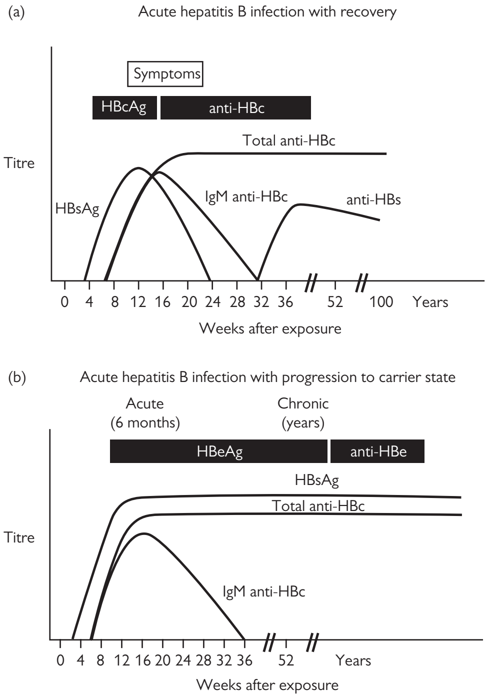

Figure 21.1 Hepatitis B serology

Source: Weinbaum CM, *et al*. *Morbidity and Mortality Weekly Report*, September 19, 2008: 57(RR08);1–20 https://www.cdc.gov/mmwr/preview/mmwrhtml/rr5708a1.htm

Management Anti-HCV antibody is detectable 3–4mo after infection; HCV-RNA tests are +ve after <2wk. Advise patients about safe sex, to avoid alcohol and not to donate organs/blood/semen. Refer to specialist services for treatment with direct-acting antiviral agents (DAAs) with a view to cure. Vaccinate against hepatitis A and B if not already immune.

Further information

BASHH 2017 interim update of the 2015 BASHH national guidelines for the management of the viral hepatitides. 🖱 www.bashhguidelines.org/media/1161/ viral-hepatitides-2017-update-18-12-17.pdf

Public Health England (2017). The green book: hepatitis B. 🖱 www.gov.uk/government/publications/hepatitis-b-the-green-book-chapter-18

Information for patients

British Liver Trust ☎ 0800 652 7330 🖱 www.britishlivertrust.org.uk

HIV infection: prevention and testing

Human immunodeficiency virus (HIV) is a retrovirus infecting T-helper cells bearing the CD4 receptor. Worldwide, the HIV epidemic continues, but prophylaxis and treatment have vastly improved prognosis in developed countries where treatment is available. It is estimated that ~1 in 3 of those infected in the UK are unaware of their diagnosis.

Transmission ~1 in 1000 exposures. Mode of transmission may be:

- Sexual (60–70%)—heterosexual intercourse in 54%
- IV drug misuse (2%)
- Mother → child (1.5%)
- Infected blood products
- Accidental (e.g. needle stick injuries)

Prevention of HIV infection[G]

- Promotion of safe sex and ↓ IV drug misuse and needle sharing
- Screening blood donors
- Prevention of transmission from mother to child—risk can be ↓ to <1% with antiretroviral treatment, elective Caesarean section, and advising against breastfeeding
- Trials of HIV vaccines are in advanced stages
- Refer to SH clinic for post-exposure prophylaxis if sexual contact <72h previously with an HIV-infected individual (or high-risk individual)
- Consider referral to SH clinic for discussion about pre-exposure prophylaxis (PrEP) for high-risk groups if available locally (e.g. HIV −ve men having condomless anal sex with men; homo- or heterosexual partners of HIV +ve individuals on treatment for <6mo or who have viral load ≥200 copies/mL)

Accidental exposure (e.g. needle stick injury) Significant exposure if: source is HIV +ve; material is blood/another infectious body fluid (semen, amniotic fluid, genital secretions, CSF), and exposure is caused by inoculation (risk 1 in 300) or by a splash onto a mucus membrane (risk 1 in 3000).

⚠ *Immediate action* Irrigate the site of exposure with running water; establish history of HIV infection and (if possible) obtain a blood sample from the source and victim. Refer to A&E immediately for HIV post-exposure prophylaxis. Treatment is with 3 antiretroviral drugs for 4wk.

HIV tests

- HIV antibodies can take 3mo to develop after infection. HIV 1/2 antibody tests can be used for screening, and diagnosis if possible exposure occurred >3mo previously. If in doubt, arrange a second test 3mo after the first
- Rapid point-of-care tests (RPOCT) are now widely available. Confirm all +ve RPOCT with a conventional blood test
- If specific risk >4wk but <3mo prior to testing, offer p24 antigen test. A –ve result excludes HIV in most—confirm –ve tests with another test after 3mo

Who should be offered an HIV test?

Universal HIV testing (offered to everyone)

- Available via the internet in some UK areas (🖱 https://freetesting.hiv/)
- Everyone attending sexual health clinics
- Women registering for antenatal care or termination of pregnancy

- Everyone diagnosed with TB, hepatitis B/C, or lymphoma
- If prevalence of HIV in the local population is >2/1000, all those registering in general practice, and all general medical admissions

High-risk patients

- If HIV is part of the differential diagnosis of the presenting condition
- If diagnosed with another STI or history of injecting drug misuse
- Sexual partners of those known to be HIV +ve
- Men who have sex with other men (MSM) and any female sexual partners
- People originating from or current/former sexual partner from a country with high HIV prevalence (>1%, e.g. sub-Saharan Africa, India)

Offer repeat testing if

- HIV −ve, but possible exposure has occurred <3mo ago, or since testing
- MSM or injecting drug users—offer >1×/y
- Women who refuse an HIV test at antenatal booking—offer testing again, then a third time at 36wk

Index conditions In which HIV is part of the differential diagnosis:

- **Lungs** TB, pneumocystis, aspergillosis, bacterial pneumonia, lung cancer
- **Nervous system** Cerebral toxoplasmosis; cerebral lymphoma; cryptococcal meningitis; progressive multifocal or other leucoencephalopathy; aseptic meningitis/encephalitis; cerebral abscess; SOL; Guillain–Barré syndrome; transverse myelitis; peripheral neuropathy; dementia
- **Skin** Kaposi's sarcoma; severe/recalcitrant seborrhoeic dermatitis or psoriasis; multidermatomal/recurrent herpes zoster
- **GI tract** Persistent cryptosporidiosis; chronic diarrhoea; hepatitis B/C; salmonella, shigella, or campylobacter infection; oral candidiasis/hairy leukoplakia; anal cancer (or intraepithelial dysplasia)
- **Sexual health** STI; cervical cancer; CIN 2/3; VIN; seminoma
- **Blood** Unexplained blood dyscrasia, e.g. thrombocytopenia, neutropenia, lymphopenia; unexplained lymphadenopathy; lymphoma
- **Eyes** CMV retinitis/infective retinal disease; unexplained retinopathy
- **Other** PUO; weight ↓; chronic parotitis/parotid cysts; head/neck cancer; mononucleosis-like syndrome (primary HIV)

Giving the result

If the result is negative Consider if the patient needs a follow-up test in 3mo. Provide information about minimizing future risk of HIV infection.

If the result is positive Give the result early in the consultation. Explain the result and its implications. Emphasize the +ve aspects of knowing the diagnosis. Try to arrange specialist referral before the patient attends. Give the patient time to talk through feelings and fears. Talk about support available, e.g. friends/family; support organizations. Provide relevant literature. Arrange follow-up to maintain the link with primary care.

Further information

BASHH (2008) UK National guidelines for HIV testing. ✆ www.bashhguidelines.org/media/1067/1838.pdf

BASHH (2015) UK guideline for the use of HIV post-exposure prophylaxis following sexual exposure. ✆ www.bashhguidelines.org/media/1027/pepse-2015.pdf

BASHH/BHIVA (2018) Guidelines on the use of HIV pre-exposure prophylaxis (PrEP). ✆ www.bashhguidelines.org/media/1189/prep-2018.pdf

HIV infection: clinical disease

Primary HIV Half have no symptoms. Possible symptoms:

- Mononucleosis-like picture of fever, fatigue, myalgia/arthralgia ± lymphadenopathy. Consider as a differential diagnosis of glandular fever
- Blotchy rash affecting the trunk, and orogenital/perianal ulceration
- Rarely acute neurological symptoms (aseptic meningitis, transverse myelitis, encephalitis) or diarrhoea
- FBC may show atypical lymphocytes
- Rarely, CD4 count drops acutely and conditions associated with immunosuppression, e.g. oral candidiasis or shingles, may occur

⚠ If you think a patient has primary HIV infection, seek urgent advice from a specialist. HIV tests can be negative <3mo after infection.

Progression Duration/severity of HIV infection is reflected by the CD4 count which ↓ as infection progresses. If CD4 count is <200 cells/mm^3, patients have acquired immune deficiency syndrome (AIDS) and are at risk from opportunistic infection (e.g. pneumococcal, TB, CMV, pneumocystis, toxoplasmosis, cryptosporidial diarrhoea) and AIDS-associated malignancies (e.g. Kaposi's sarcoma, lymphoma)—Table 21.1.

Kaposi's sarcoma Presents as purple papules/plaques on skin or mucosa of any organ. Metastasizes to lymph nodes. If suspected refer for expert help:

- Endemic Occurs in central Africa. Peripheral lesions, good response to chemotherapy
- Associated with AIDS or transplant Commonly skin or pulmonary lesions; lymphatic obstruction predisposes to cellulitis

Lymphogranuloma venereum (LGV) Caused by *Chlamydia trachomatis* infection; almost exclusively affects HIV +ve men who have sex with men. Primary lesion is a small papule (easily missed). The secondary phase of the disease follows after 10–30d and results in enlarged inguinal ± femoral LNs—usually unilateral (>60%). May ulcerate and form fistulae. If suspected, refer to a SH clinic. Treatment is with doxycycline 100mg bd po for 3wk.

Psychological support Provide information about local and national support organizations to all at diagnosis and subsequently if needed.

Antiretroviral drugs (ARDs) *Options*:

- Nucleoside reverse transcriptase inhibitors (NRTIs), e.g. zidovudine
- Non-nucleoside reverse transcriptase inhibitors (NNRTIs), e.g. efavirenz
- Protease inhibitors, e.g. ritonavir—associated with facial lipodystrophy with long-term use, and multiple drug interactions
- Entry inhibitors—inhibit the fusion of HIV to the host cell, e.g. enfuvirtide
- HIV integrase inhibitors, e.g. dolutegravir
- Cobicistat boosts effectiveness of other ARTs but no antiviral activity itself

Choice of drugs is a specialist decision. A combination is usual (known as 'highly active anti-retroviral therapy' or HAART). Adherence to therapy is essential to avoid resistance. The aim is to ↓ viral load to an undetectable level in <6mo. Response to treatment is measured with viral load, CD4 count, and CD4 percentage. Treatment failure requires switching or ↑ therapy. ❗ Do not stop/change dose of ARDs without taking specialist advice.

Table 21.1 CD4 counts and HIV-related problems

CD4 count (cells/mm^3)	Risk of opportunistic infection	Risk of HIV associated tumours
>500	Minimal/none	Very small ↑ risk
200–500	Little risk unless falling rapidly, except TB	Small ↑ risk
<200	↑ risk of serious opportunistic infection, e.g. • Pneumocystis pneumonia • Toxoplasmosis • Oesophageal candidiasis	Increasing risk
<100	Additional risk of: • *Mycobacterium avium intracellulare* • Cytomegalovirus	High-risk and increasingly aggressive disease

Drug interactions Record ARDs on GP records even though usually supplied by specialists. Check interactions at: 🖱 www.hiv-druginteractions.org

Immune reconstitution syndrome ↑ in immune function with ARD treatment may → an inflammatory reaction to opportunistic organisms in the first weeks/months. Autoimmune disorders (e.g. vitiligo) may also be triggered.

Prophylaxis against opportunistic infections Prophylactic antibiotics are used to prevent pneumocystis, toxoplasmosis, and *Mycobacterium avium* for patients with low CD4 counts.

Immunizations

- Inactivated vaccines Can be used safely; offer annual influenza vaccination, pneumococcal vaccination, and hepatitis A/B vaccination if seronegative
- Live vaccines (e.g. BCG) Generally contraindicated but give varicella (for all) and MMR (♀ of childbearing age) if seronegative and CD4 count $>200mm^3$

CVD prevention HIV is associated with ↑ risk of CVD. Check risk before, then 3–6mo after starting ARDs and annually thereafter (every 5y from 40y if not taking ARDs). Provide lifestyle advice (➲ p. 214), and treat hyperlipidaemia if 10y CVD risk >10% (➲ p. 222).

Sexual health Advise to use condoms with water-based lubricant for all vaginal/anal sex and condoms/dams for oral sex—even if both partners are HIV +ve (as prevents transmission of drug-resistant strains).

- Contraception Efficacy of hormonal contraception is ↓ by antiretrovirals. Consider depot medroxyprogesterone or IUS/Cu-IUD. Advise condoms in addition to contraception. If emergency contraception is needed, first-line is an IUD; if using levonorgestrel, provide a double (3mg) dose—➲ p. 726
- Cervical screening Women with HIV have ↑ risk of HPV and cervical cancer. Annual cervical screening is recommended

Contraception ➲ p. 729 **Pregnancy** ➲ p. 797

Palliative care ➲ p. 1011

Death Record on death certificates if HIV has contributed to death.

Information and support for patients with HIV and carers

NAM Aidsmap 🖱 www.aidsmap.com
National AIDS Trust 🖱 www.nat.org.uk
Terrence Higgins Trust ☎ 0808 802 1221 🖱 www.tht.org.uk

Other sexually transmitted infections

Genital herpes[G] HSV is transmitted by direct contact with lesions. Lesions may appear anywhere on the skin or mucosa but are most frequent around the mouth, on the lips, conjunctiva, cornea, and genitalia.

Presentation of primary infection May be asymptomatic. If symptomatic, history and examination are diagnostic in 90% cases. Presents with multiple painful genital ulcers on a red background ± inguinal lymphadenopathy <1wk after sexual contact. Lesions crust over then heal. Untreated lasts 3–4wk. *Complications*: urinary retention, aseptic meningitis.

Management

- Refer to a SH clinic if diagnosis is uncertain and for contact tracing
- Treat with aciclovir if presents ≤5d after symptoms start and while new lesions are still forming (↓ duration, symptoms, and complications)
- Analgesia, ice packs, and salt baths may help. 5% lidocaine ointment gives symptom relief but use with caution as may cause sensitization
- Advice—barrier methods of contraception (risk of transmission in monogamous relationships is 10%/y)
- If pregnant obtain specialist advice

Recurrent infection Reactivation of latent virus. Less severe than primary infection. Neonatal transmission rates are low (<3%)—elective Caesarean section for those with active recurrences at term is controversial. Consider suppressive therapy if 6 attacks/y, e.g. aciclovir 400mg bd. If breakthrough occurs, ↑ dose of antiviral, e.g. aciclovir 400mg tds.

> *Neonatal infection* Presents at age 5–21d with vesicular lesions around the presenting part or rarely systemic infection. Usually babies of women with no history of genital HSV. Refer as a paediatric emergency.

Genital warts[G] Caused by human papillomavirus (HPV). Usually sexually transmitted, and >25% have other concomitant STIs. Disease may be clinical (found on examination) or subclinical (changes associated with infection detected on smear). In women, CIN (➔ p. 702) is related to infection with HPV types 16 and 18, but 90% of genital warts are caused by HPV types 6 or 11. Prevalence is ↓ since introduction of routine HPV vaccination.

Presentation in women Often asymptomatic but may be associated with itching or vaginal discharge. Warts are usually seen on the vulva or introitus. Warts enlarge during pregnancy.

Presentation in men Usually found on the penis/perianally. ❗ Perianal warts do not always indicate anal sex—avoid assumptions about sexuality.

Management of clinical warts Treatment does not eradicate the virus but removes lesions. Barrier contraception is needed for at least 3mo after the warts are gone. Treatment options in primary care:

- Podophyllotoxin Suitable for home treatment of unkeratinized genital warts and licensed for a 4wk course. Avoid in pregnancy. Apply 2×/d for 3d, followed by a 4d rest. This cycle is repeated 4–5×. Side effects: soreness/ulceration of the genital skin—advise to discontinue treatment
- Imiquimod Can be used at home for keratinized or non-keratinized warts. Avoid in pregnancy. Apply 3×/wk. On each occasion wash off

6–10h later. Continue for up to 16wk. Avoid unprotected intercourse after application. Can weaken latex condoms

Alternatives (usually in specialist settings) Trichloroacetic acid, excision, cryotherapy, electrosurgery.

> *Human papillomavirus (HPV) vaccination* is aimed at preventing infection with strains causing cervical cancer. Currently vaccines target strains 6, 11, 16, and 18, which account for ~70% of HPV-related cancer cases and >90% of genital warts. Vaccination in the UK is targeted at girls aged 12–14y. Cervical screening in adulthood is still necessary as the vaccine does not protect against all strains causing cervical cancer.

Pubic liceG Pubic (or crab) lice are similar to head lice and may be sexually transmitted. All hairy areas (including eyelashes, eyebrows, pubic and axillary hair) can be affected. Repeat treatment after 3–7d. Treatment options are:

- Malathion 0.5%—apply to dry hair; wash out after 12h
- Permethrin 1% cream rinse—apply to damp hair; wash out after 10min
- Phenothrin 0.2%—apply to dry hair; wash out after 2h
- Carbaryl 0.5–1% (unlicensed)—apply to dry hair; wash out after 12h

Scabies ➔ p. 613 **Molluscum contagiosum** ➔ p. 609

SyphilisG Caused by *Treponema pallidum*. Rare in the UK but incidence is increasing. *Incubation*: 9–90d. If suspected, send blood for VDRL, TPHA or treponemal antibody absorption depending on local policy. In all confirmed cases refer for specialist care. Contact tracing is essential. *4 stages*:

- Primary syphilis Chancre at the site of contact
- Secondary syphilis 4–8wk after chancre—systemic symptoms: fever, malaise, generalized lymphadenopathy, anal papules (condylomata lata), rash (trunk, palms, soles), buccal snail track ulcers, alopecia
- Tertiary syphilis 2–20y after initial infection—gummas (granulomas) in connective tissue, e.g. testicular gumma
- Quaternary syphilis Cardiovascular or neurological complications

Further information

BASHH (2008) United Kingdom national guideline on the management of *Phthirus pubis* infestation. 🖱 www.bashhguidelines.org/media/1074/28.pdf

BASHH 2014 UK national guideline for the management of anogenital herpes. 🖱 www.bashhguidelines.org/media/1019/hsv_2014-ijstda.pdf

BASHH 2016 UK national guideline on the management of scabies. 🖱 www.bashhguidelines.org/media/1137/scabies-2016.pdf

BASHH UK national guidelines on the management of syphilis 2015. 🖱 www.bashhguidelines.org/media/1148/uk-syphilis-guidelines-2015.pdf

BASHH UK National guidelines on the management of anogenital warts 2015. 🖱 www.bashhguidelines.org/media/1075/uk-national-guideline-on-warts-2015-final.pdf

Information for patients

Herpes Association ☎ 0845 123 2305 🖱 www.herpes.org.uk
National Sexual Health Helpline ☎ 0300 123 7123

Contraceptive choices

80% women receive contraceptive advice and treatment through their GP. A sexually active woman has an 85% chance of becoming pregnant in <1y without contraception, and ~1 in 3 pregnancies are unplanned.

Contraceptive services Provided by GP practices as an *Additional Service*; opting out results in a 2.4% ↓ in global sum. A payment may be available for GPs to fit IUDs according to local contracts.

Emergency contraception (EC)[G]

Earliest likely ovulation date = date of first day of LMP + (length of shortest cycle in days − 14). Cycles must be regular and LMP accurate.

Copper IUD (Cu-IUD) Offer as first line. Can be inserted ≤5d after earliest likely date of ovulation (i.e. up to day 19 of a regular 28d cycle) or, if after day 19, ≤120h (5d) after unprotected sexual intercourse (UPSI) if that is the only episode of UPSI in the cycle. >99% effective; provides ongoing contraception. Progestogen-containing IUDs are not suitable for this purpose.

Oral EC Delays ovulation. Ineffective once ovulation has occurred. 2 types—both are available OTC or on prescription:
- Ulipristal acetate (UPA) 30mg po stat. Licensed for use 1×/cycle only ≤120h (5d) after UPSI. Delay ≥5d before starting regular contraception
- Levonorgestrel 1.5mg po stat. Licensed ≤72h (3d) after UPSI but effective for ~96h (4d). Can be used ≥1×/cycle. Half as effective as UPA

Possible pitfalls of oral EC
- Vomiting <3h after oral dose—give replacement dose or offer Cu-IUD
- Enzyme-inducing drugs (e.g. antiepileptics), BMI >26kg/m^2 or weight >70kg ↓ efficacy of oral preparations. Consider a Cu-IUD or ↑ dose of levonorgestrel to 3mg (unlicensed). ❗ Do not ↑ the dose of UPA

All women Provide advice about ongoing contraception. Advise to return if abdominal pain, or next period is overdue or abnormally light/heavy.

Before providing routine contraception Provide information about contraceptive options (Table 21.2) and written information on chosen method(s). Exclude pregnancy. Discuss prevention of STIs. Advise high-risk groups to use barrier methods in addition to other methods of contraception.

Choice of method See Table 21.2. Consider: UK Medical Eligibility Criteria (UK MEC); preference; age; lifestyle/cultural aspects; medical history; and risk of STI. ❗ Contraceptives are free of charge for all women in the UK.

Excluding pregnancy No symptoms/signs of pregnancy and ≥1 of:
- No intercourse, or <5d since the start of LMP, childbirth, abortion, miscarriage, ectopic pregnancy or uterine evacuation for trophoblastic disease
- Correctly using a reliable method of contraception
- No intercourse for >21d *and* has had a negative high-sensitivity urine pregnancy test (able to detect hCG levels around 20 mIU/mL)
- <21d postpartum (non-breastfeeding women)
- Fully breastfeeding, amenorrhoeic, and <6mo postpartum

Table 21.2 Summary of contraceptive methods

Method of contraception	% of unintended pregnancies[a]	Advantages	Disadvantages
Sterilization (♂) ➔ p. 744	0.05	No contraindications Single procedure	Difficult to reverse Postoperative complications
Sterilization (♀) ➔ p. 744	0.5	No contraindications Single procedure	Requires general anaesthetic Difficult to reverse Postoperative complications ↑ risk ectopic pregnancy
Nexplanon® ➔ p. 739	0.05	Lasts 3y Immediately reversible	Training needed to insert/remove Wound infection and/or scarring Can cause irregular bleeding Progestogenic side effects
Progestogen-containing intrauterine system (IUS) ➔ p. 740	0.1	Lasts 5y Immediately reversible ↓ bleeding and dysmenorrhoea ↓ ectopic pregnancy Endometrial protection	Training needed for insertion May cause erratic bleeding Progestogenic side effects Problems with insertion/retrieval
Copper intrauterine device (Cu-IUD) ➔ p. 740	0.1	Lasts ≥5y Immediately reversible No systemic effects	Training needed for insertion Heavy periods No protection from ectopic pregnancy Problems with insertion/retrieval
Combined contraceptive pill/patch/ring ➔ p. 730	**0.3** (9)	Regular cycle and lighter periods ↓ dysmenorrhoea Cycle control	Poor compliance Side effects ↑ risk breast cancer and thromboembolism
Progestogen-only pill (POP) ➔ p. 736	**0.3** (8)	Few side effects/contraindications Can abolish periods	Poor compliance Irregular bleeding Progestogenic side effects
Injectable progestogen ➔ p. 737	**0.3** (3)	Avoids pill-taking ↓ bleeding and can help PMS ↓ risk of ectopic pregnancy and endometrial cancer	Menstrual irregularity Weight gain Unpredictable return of fertility ↑ risk of osteoporosis
Condoms ➔ p. 744	**2** (32)	Barrier to transmission of STIs	User dependent Allergy High failure rate
Natural methods ➔ p. 745	**1** (27)	No contraindications or side effects	Teaching required High failure rate

[a] Failure rates stated are with perfect use. Rates in brackets are with typical use

Further information

FSRH (2016) UKMEC. 🖱 www.fsrh.org/ukmec

FSRH (2017) Emergency contraception. 🖱 www.fsrh.org/documents/ceu-clinical-guidance-emergency-contraception-march-2017

Box 21.2 Enzyme-inducing antiepileptic medications

- Carbamazepine
- Eslicarbazepine acetate
- Oxcarbazepine
- Phenobarbital
- Phenytoin
- Primidone
- Rufinamide
- Topiramate (if dose >200mg)
- Perampanel

Quick starting^{G} Contraception is usually initiated at the start of a menstrual cycle when it can be sure the woman is not pregnant. With quick starting, contraception is started at any time during the cycle. This ↓ risk of unintended pregnancy. Follow the same procedure as usual but advise about the need for additional barrier contraception or abstinence until the chosen method becomes effective (Table 21.3).

If pregnancy cannot be excluded Only quick start if a woman is at risk of pregnancy from further UPSI and/or wants to start contraception immediately. Follow-up with a pregnancy test after 21d. ❗ There is no evidence that fetal exposure to hormonal contraceptives is harmful.

Quick starting after emergency contraception CHC, POP, and progestogen implant are suitable. Avoid injectable progestogen as irreversible and limited safety data in pregnancy. Consider quick starting other 'bridging' contraception if chosen contraceptive cannot be started immediately. After UPA, delay quick starting for ≥120h (5d). Advise about barrier contraception until quick start method becomes effective (Table 21.3).

Special groups

- Teenagers and women >40y—➔ p. 746
- Postpartum—➔ p. 816

Breast cancer or history of breast cancer Use non-hormonal forms of contraception e.g. barrier methods, Cu-IUD or sterilization.

Inflammatory bowel disease^{G} All contraceptive methods are suitable for women with inflammatory bowel disease (IBD). Special considerations:

- **Efficacy of oral contraception** May be ↓ if malabsorption due to severe small bowel disease or resection, or vomiting/severe diarrhoea for >24h
- **Long-term use of injectable progestogens** Both IBD and long-acting injectable progestogens are associated with ↑ risk of osteoporosis. Weigh risks/benefits of use and reassess every 2y.

Epilepsy Efficacy of oral hormonal contraception and contraceptive implants can be ↓ by antiepileptic medications that ↑ the activity of hepatic cytochrome enzyme P450. For women taking enzyme-inducing antiepileptic medication (Box 21.2), intrauterine contraception (IUS or Cu-IUD) or long-acting injectable progestogens are contraceptives of choice. If a woman wishes to use combined oral contraceptives, efficacy can be ↑ by:

- Increasing oestrogen dose to 50–70 micrograms/d
- Tri-cycling—taking 3 packets back to back without a pill-free interval, and
- ↓ the pill-free interval to 4d

Alternatively advise using a barrier as well as a hormonal method.

⚠ Avoid prescribing sodium valproate to women/girls of childbearing age. If essential, ensure a pregnancy prevention plan is in place and that information is provided about potential teratogenic effects both verbally and in writing. Lamotrigine is not an enzyme-inducing drug but combined hormonal contraceptives ↓ efficacy of lamotrigine → poor seizure control.

Table 21.3 Need for additional contraception after quick starting

	Additional contraception required	
Day of the menstrual cycle	*1–5*	*6 onwards*
Combined hormonal contraception[a]	None	7d
Progestogen-only pill	None	2d
Progestogen-only injection or implant	None	7d
Day of the menstrual cycle	*1–7*	*8 onwards*
IUS	None	7d
Cu-IUD	None	None

[a] If started on any day other than the first day of the woman's period, recommend additional contraception for 7d (Zoely®) and 9d (Qlaira®)

HIV[G] Dual protection using barrier *and* hormonal/intrauterine contraception prevents both pregnancy and ↓ HIV transmission (although condoms are not always required if on suppressive ART). *Special considerations*:

- CD4 count <200 cell/mm^3 Intrauterine contraceptive devices (IUS and Cu-IUD) are contraindicated due to ↑ risk of infection during insertion; if already *in situ*, devices can be left in place
- Anti-retroviral therapy (ART) Efficacy of oral hormonal contraception and contraceptive implants can be ↓ by enzyme-inducing ART. Consider altering ART regimen or another method of contraception
- Condoms containing spermicide Nonoxinol-9 (N-9) causes mucosal irritation and can ↑ transmission rates of HIV. Avoid use
- Long-term use of injectable progestogens Both HIV and long-acting injectable progestogens are associated with ↑ risk of osteoporosis. Weigh risks/benefits of use and reassess every 2y

Cardiac disease[G] Choices can be restricted:

- Combined hormonal contraception Associated with ↑ risk of venous thromboembolism. Contraindicated if significant cardiac disease. Only consider if low risk (i.e. discharged from cardiology follow-up or seen at intervals of >2y; normal peripheral oxygen saturations; and not taking any long-term cardiac medication including aspirin)
- Progestogen-only pills, injections, and implants Generally suitable but efficacy of implants/pills may be ↓ if concurrently taking enzyme-inducing drugs
- Intrauterine contraceptives May be suitable, but take cardiology advice if at high risk of bacterial endocarditis (➔ p. 244), or history of arrhythmia or congenital heart disease (may need to be fitted in a hospital setting)

Further information

BHIVA/BASHH/FSRH (2017) BHIVA/BASHH/FSRH guidelines for the sexual & reproductive health of people living with HIV. 🖱 www.bhiva.org/file/zryuNVwnXcxMC/SRH-guidelines-for-consultation-2017.pdf

FSRH (2014) Contraceptive choices for women with cardiac disease. 🖱 www.fsrh.org/standards-and-guidance/documents/ceu-guidance-contraceptive-choices-for-women-with-cardiac

FRSH (2016) Sexual and reproductive health for individuals with inflammatory bowel disease. 🖱 www.fsrh.org/standards-and-guidance/external/ceu-clinical-guidance-srh-ibd-digital-version

FSRH (2017) Quick starting contraception. 🖱 www.fsrh.org/standards-and-guidance/current-clinical-guidance/quick-starting-contraception/

Combined hormonal contraception

Contraceptives containing an oestrogen and progestogen are available as:

- Combined oral contraceptive pills (COC)
- Contraceptive patches (Evra®)
- Vaginal contraceptive rings (NuvaRing®)

COC pill Most COC come in packets of 21 pills. The woman takes the entire packet, starting on the first day of her cycle and then has a 7d 'pill-free' break before starting the next packet. Pills vary by:

Oestrogen type Most COCs contain ethinylestradiol; alternative is estradiol valerate (Qlaira®).

Oestrogen content

- Low-strength preparations (20 micrograms ethinylestradiol) Use if aged >40y, risk factors for circulatory disease, continuous regimen, or oestrogenic side effects
- Standard-strength preparations (30–35 micrograms ethinylestradiol) Use for most women
- Phased preparations Dose of oestrogen/progestogen varies through the cycle. Useful if bleeding problems with monophasic products
- Everyday (ED) preparations Taken continuously and include 7 inert pills. Can help women who find it difficult to remember to start a new packet

Progestogen type COC pills containing:

- Levonorgestrel Suitable for most women. Choose for first-time COC pill users
- Desogestrel, norgestimate, dienogest, drospirenone, and gestodene Consider if side effects, e.g. acne, headache, depression, weight ↑, breast symptoms, breakthrough bleeding. ❗ Desogestrel/gestodene may be associated with ↑ clotting risk
- Cyproterone acetate (co-cyprindiol) Licensed for treatment of acne, not for contraception, but does provide contraception. Use for 3mo after acne control is achieved. Associated with 4× ↑ risk of venous thromboembolism compared to COC containing levonorgestrel

Contraceptive patch (Evra®) 20 micrograms ethinylestradiol and norelgestromin in a transdermal patch. Apply patch on day 1 of the cycle; change patch on days 8 and 15; remove third patch on day 22 and then apply new patch after a 7d 'patch-free' interval to start the subsequent cycle.

Contraceptive vaginal ring 15 micrograms/24h ethinylestradiol and etonogestrel. Alternative low-dose preparation. Insert ring into vagina on day 1 of cycle and leave in for 3wk; remove ring on day 22; subsequent courses repeated after 7d ring-free interval.

Reasons to avoid combined contraception Box 21.3

Further information

FRSH (2019) Combined hormonal contraception. 🖱 www.fsrh.org/standards-and-guidance/documents/combined-hormonal-contraception
FSRH (2016) UK MEC 🖱 www.fsrh.org/ukmec

Box 21.3 Reasons to avoid combined hormonal contraception*

Venous disease

- Avoid if sclerosing treatment for varicose veins or if history of current/past venous thromboembolism (VTE)
- Risk factors for VTE (use with caution if 1; avoid if >1):
  - Age ≥35y—avoid if ≥50y
  - Smoker or <1y after smoking cessation—avoid if ≥35y and if smoking
  - BMI ≥30kg/m^2—avoid if BMI ≥35kg/m^2
  - Family history of VTE in first-degree relative <45y—avoid if known prothrombotic coagulation abnormality, e.g. factor V Leiden, antiphospholipid antibodies, lupus anticoagulant
  - Immobility—avoid if bed-bound or leg in plaster cast
  - History of superficial thrombophlebitis

Arterial disease

- Avoid if valvular/congenital heart disease with history of complications (e.g. pulmonary hypertension, AF, SBE) or if history of CVD including stroke/TIA, IHD, peripheral vascular disease, hypertensive retinopathy
- Risk factors for CVD (use with caution if 1; avoid if >1):
  - Age ≥35y—avoid if ≥50y
  - Smoker—avoid if smoking ≥40 cigarettes/d
  - BMI ≥30kg/m^2—avoid if BMI ≥35kg/m^2
  - Family history of arterial disease in first-degree relative <45y—avoid if atherogenic lipid profile
  - DM—avoid if vascular, renal, neurological, or eye complications
  - Hypertension with BP >140/90mmHg—avoid if >160/95mmHg
  - Migraine without aura—avoid if migraine with aura, or migraine without aura starts or worsens after starting CHC

Liver disease Avoid if active/flare of viral hepatitis, liver tumour, or severe cirrhosis or if active gall bladder disease; seek specialist advice if history of contraceptive-associated cholestasis.

Cancer Avoid if current breast cancer; take specialist advice if no suitable alternative and past history of breast cancer, but no evidence of disease for >5y or known gene mutation for breast cancer (e.g. *BRCA1/2*).

Pregnancy-related issues Avoid if history in pregnancy of pruritus, cholestatic jaundice, chorea, or pemphigoid gestationis; or if <21d postpartum (not breastfeeding) or <6wk postpartum (breastfeeding)—➔ p. 816.

Drug interactions ➔ p. 734

Others Avoid if acute porphyria or haemolytic uraemic syndrome.

❗ Investigate any undiagnosed vaginal bleeding before starting combined hormonal contraception.

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* Based on UK Medical Eligibility Criteria (UKMEC) 3 (theoretical or proven risks usually outweigh advantages) and 4 (unacceptable health risk).

Before starting combined contraception

- Take a history—medical, sexual health, medications, and lifestyle
- Consider asking for specialist haematology advice about thrombophilia screening if FH of DVT/PE in a first-degree relative aged <45y or multiple family members, and/or check cholesterol/triglycerides if FH of arterial disease in a first-degree relative <45y, or multiple family members
- Check BP
- Education—use UKMEC to discuss side effects/risks of combined contraception (Table 21.4). In particular, warn to stop CHC and seek medical advice if any symptoms/signs of DVT. Advise about STIs, cervical smears, smoking, control of weight. Give both verbal and written directions on use

Starting combined contraception Contraceptive effect starts immediately if started:

- Day 1–5 of the cycle (day 1 only for oestradiol valerate/dienogest pill)
- At the end of the third week postpartum
- <5d after miscarriage/TOP at <20wk gestation (day 1 only for estradiol valerate/dienogest pill)
- Changing COC pill variety or to patch or ring—start the new pill/patch/ring omitting the 7d break (or 'inactive' tablets if taking ED preparation)
- Changing from contraceptive implant, injectable progestogen, or desogestrel-only POP—start at any time until repeat injection is due, implant is due for removal, or last desogestrel pill taken

❗ In all other cases, and if quick starting combined contraception at another point in the cycle use additional contraception for the first 7d of use (9d if using estradiol valerate/dienogest pill).

Extended dosing[G] Continuous dosing is an alternative approach to combined hormonal contraceptive administration that does not ↓ contraceptive efficacy. Several (unlicensed) regimens are in common use:

- Short pill-free interval Replacement of 7d break with 4d break
- Tri-cycling 3 cycles taken continuously back-to-back followed by a 7d break, i.e. 3 × 21 monophasic COC, 3 × rings, or 9 × patches
- Extended use Continuous use of monophasic COC, ring, or patch until breakthrough bleeding for 3–4d, followed by a 4d or 7d break

Follow-up 3mo after starting or changing a combined contraceptive—earlier if complications. Once established, review every 6–12mo. At follow-up, assess risk factors and side effects; give health education, e.g. smoking cessation advice, benefits of long-acting reversible contraception, information about STIs; check BP.

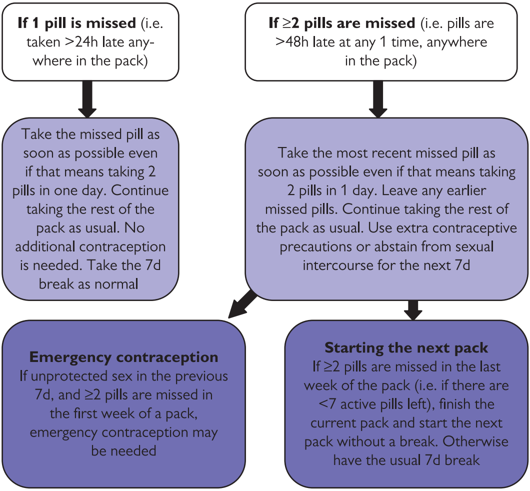
Table 21.4 Risks and benefits of combined hormonal contraception use

Risks	Benefits
Venous thromboembolism (risk ↑ ×2 but absolute risk is still very low) Ischaemic stroke—small ↑ risk Breast and cervical cancer—any ↑ in risk is small and disappears <10y after combined contraception is stopped Mood changes (but no ↑ in depression)	Improvement in acne ↓ in menstrual pain and bleeding ↓ in symptoms of PMS (➲ p. 680) ↓ in menopausal symptoms ↓ risk of ovarian, bowel, and endometrial cancer that persists after combined contraception has stopped No evidence of weight ↑

Missed doses COC pills (except Qlaira®)—Figure 21.2; missed Qlaira® or contraceptive patches/rings—see *BNF* and/or product literature.

⚠ **Reasons to stop CHC immediately** (pending investigation if needed)

- Sudden severe chest pain (even if not radiating to left arm)
- Sudden breathlessness (or cough with bloodstained sputum)
- Unexplained swelling or severe pain in calf of one leg
- Acute abdominal pain
- Serious neurological effects including:
  - Unusual severe, prolonged headache especially if first time or getting progressively worse
  - Sudden dysphasia, partial or complete loss of vision, disturbance of hearing, or other perceptual disorders
  - Bad fainting attack or unexplained collapse
  - First unexplained epileptic seizure
  - Weakness, motor disturbances, or numbness affecting one side or one part of body
- Hepatitis, jaundice, liver enlargement
- BP >160/95mmHg
- Prolonged immobility after surgery or leg injury
- Detection of a risk factor/contraindication (➔ p. 731)



If 1 pill is missed (i.e. taken >24h late anywhere in the pack)

Take the missed pill as soon as possible even if that means taking 2 pills in one day. Continue taking the rest of the pack as usual. No additional contraception is needed. Take the 7d break as normal

If ≥2 pills are missed (i.e. pills are >48h late at any 1 time, anywhere in the pack)

Take the most recent missed pill as soon as possible even if that means taking 2 pills in 1 day. Leave any earlier missed pills. Continue taking the rest of the pack as usual. Use extra contraceptive precautions or abstain from sexual intercourse for the next 7d

Emergency contraception
If unprotected sex in the previous 7d, and ≥2 pills are missed in the first week of a pack, emergency contraception may be needed

Starting the next pack
If ≥2 pills are missed in the last week of the pack (i.e. if there are <7 active pills left), finish the current pack and start the next pack without a break. Otherwise have the usual 7d break

Figure 21.2 Advice for patients regarding missed COC pills

Further information

FRSH (2019) Combined hormonal contraception. 🖱 www.fsrh.org/standards-and-guidance/documents/combined-hormonal-contraception

Short-term side effects Usually resolve within 2–3 cycles.

Relative oestrogen excess Breast tenderness (3.6%); nausea (1.5%); dizziness; cyclical weight ↑; bloating; vaginal discharge without infection. Use a more progestogen-dominant pill.

Relative progestogen excess Mood swings (3.9%); PMT; dry vagina; sustained weight ↑; ↓ libido; lassitude; acne. Use a more oestrogen-dominant pill.

Headache Affects 2.9% of women taking the combined contraceptive. Ask women to report ↑ in headache frequency or onset of focal symptoms when taking any combined contraceptive. If new focal symptoms, discontinue immediately and, if not typical of migraine aura and lasts >1h, admit. If headaches continue consider switching brand/alternative method of contraception.

Breakthrough bleeding Most common in the first few months of combined contraceptive use—after 6 cycles affects 1.1% women (spotting affects 3.3% women). If no vomiting/diarrhoea and no missed pills, breakthrough bleeding does not indicate ↓ efficacy. If symptoms suggest other pathology (e.g. abdominal or pelvic pain, post-coital bleeding) or breakthrough bleeding persists >3mo:

- Check compliance—any missed pills? Breakthrough bleeding may start 2–3d after a missed pill; any diarrhoea/vomiting
- Check for gynaecological causes—exclude STI (especially chlamydia); examine cervix; check smear is up to date and take smear if overdue; exclude pregnancy; consider referral for ultrasound, hysteroscopy, + endometrial sampling if >45y or other risk factors for endometrial cancer

↑ oestrogen content of COC pill if on low-dose preparation. If problem persists, change progestogen. If still persists, ↑ progestogen and/or try phased preparation.

Long-term risks/benefits See Table 21.4, ➔ p. 732.

Acne and CHC In general acne improves when using CHC. If it fails to improve, consider switching to a brand containing a less androgenic progestogen (e.g. desogestrel, drospirenone) or one with a higher oestrogen content. Co-cyprindiol use is associated with higher risk of thromboembolism; if using for contraception as well as acne control, switch to an alternative CHC 3mo after control of acne symptoms has been achieved.

Hepatic enzyme-inducing drugs Combined contraceptives may interact with hepatic enzyme-inducing drugs leading to ↓ efficacy, e.g.:

- Anti-infective agents—rifamycins (rifampicin, rifabutin), griseofulvin, antivirals (e.g. nelfinavir, nevirapine, ritonavir)
- St John's wort
- Anticonvulsants—phenytoin, carbamazepine, oxcarbazepine, phenobarbital, primidone, topiramate, modafinil

Short course (<7d) of enzyme-inducing drug Advise additional barrier contraception while taking the enzyme-inducing drug and for 4wk after stopping it. Omit pill/patch-free week or inactive tablets if using an 'ED' preparation.

Longer course of enzyme-inducing drug Advise another unaffected method of contraception, e.g. intrauterine device. Refer for specialist advice if other methods of contraception are unacceptable to the patient.

❗ There is no evidence that broad-spectrum antibiotics (e.g. amoxicillin) ↓ efficacy of combined contraceptives. Additional contraceptive precautions are no longer recommended.

Anticonvulsants that do not affect pill efficacy

- Sodium valproate
- Lamotrigine—but seizure frequency may ↑ when combined contraception and lamotrigine are used together and side effects of lamotrigine may be ↑ when combined contraception is stopped

Interaction with ulipristal acetate (UPA) UPA blocks the action of progesterone and so ↓ effectiveness of combined contraceptives. Do not use UPA as emergency contraception if CHC has been used in the previous 7d. Do not start CHC for 5d after UPA use and then advise abstinence/additional contraception (e.g. with condoms) for a further 7d.

Diarrhoea and vomiting Does *not* affect the contraceptive patch or ring. If a woman vomits <2h after taking a COC pill or has very severe diarrhoea, assume the COC pill has not been absorbed and treat as a missed pill (➔ p. 733).

Surgery Combined contraceptives should be discontinued and alternative contraceptive arrangements made (e.g. depot injection, barrier methods) 4wk before major elective surgery, all surgery to the legs, or surgery which involves prolonged immobilization of a lower limb. Restart the combined contraceptive on the first day of the next period occurring ≥2wk after full mobilization.

Long journeys and DVT Women taking combined contraceptives are at ↑ risk of DVT during travel involving long periods of immobility (>3h). Advise women:

- To drink plenty of non-alcoholic fluids
- To keep their legs moving while sitting, or walk up and down the aisle

Graduated compression hosiery is available for purchase OTC and does ↓ risk of DVT.

Further information

BNF Treatment summary: contraceptives, hormonal. 🖱 https://bnf.nice.org.uk/treatment-summary/contraceptives-hormonal.html
FSRH (2016) UKMEC. 🖱 www.fsrh.org/ukmec
FRSH (2019) Combined hormonal contraception. 🖱 www.fsrh.org/standards-and-guidance/documents/combined-hormonal-contraception

Progestogen-only contraception

Progestogen-only contraceptives thicken cervical mucus, ↓ endometrial receptivity, and some methods inhibit ovulation. They ↓ risk of pelvic infection and can be used when oestrogen is contraindicated.

Reasons to avoid progestogen-only contraception

- Current breast cancer—may be used with specialist advice if disease free for >5y and no other suitable method of contraception
- Trophoblastic disease—seek specialist advice if unsure
- Liver disease—active viral hepatitis; severe decompensated cirrhosis, or liver tumour (benign or malignant)
- If new symptoms/diagnosis of ischaemic heart disease, stroke/TIA, or migraine with aura when taking progestogen-only contraception

❗ Investigate any undiagnosed vaginal bleeding before starting progestogen-only contraception.

Progestogen-releasing intrauterine system (IUS) ➔ p. 740

Progestogen-only pill (POP or 'mini-pill') Oral POPs are a suitable alternative for women for whom COC pills are contraindicated, e.g.:

- Women aged >50y
- Smokers aged >35y
- Women with past history/predisposition to venous thromboembolism
- Patients with ↑ BP, valvular heart disease, DM, or migraine with aura

Choice of POP 3 types are currently available in the UK:

- Norethisterone 350 micrograms daily
- Levonorgestrel 30 micrograms daily
- Desogestrel 75 micrograms daily (Cerazette®)—use if compliance problems (12h window before 'missed pill'), history of ectopic pregnancy or ovarian cysts (desogestrel POPs have a stronger ovarian suppressive effect than other POPs), and/or weight >70kg

Side effects

- Traditional POPs (not desogestrel) have higher failure rate than COC pills
- Menstrual irregularities—oligomenorrhoea, menorrhagia, amenorrhoea—examine to exclude a pathological cause ± do a pregnancy test. Menstrual irregularities tend to resolve with long-term use. If necessary, consider changing progestogen or ↑ to 2 pills/d (unlicensed)
- ↑ risk of ectopic pregnancy. If a patient presents with abdominal pain treat as an ectopic pregnancy (➔ p. 768 until proven otherwise
- Others—nausea/vomiting; headache; dizziness; breast discomfort; depression; skin disorders; disturbance of appetite; weight changes; ↓ libido
- Long term—small ↑ risk breast cancer; risk reverts to normal <10y after stopping the POP

Starting the POP

- **No previous hormonal contraception** Start on day 1–5 of the cycle—no additional contraception needed; if starting any other time, use additional contraception/abstain from sexual intercourse for 2d (if quick started after emergency contraception using UPA, wait 5d to start and then use condoms/abstinence for 2d). ❗ Do not use UPA as emergency contraception if POP taken in the previous 7d

- **Changing from COC** Start the day following completion of COC without a break (omitting 'inactive' pills if ED preparation)—no additional contraception needed
- **Changing from IUD** If POP started 2d before removal of copper IUD or at the time of IUS removal—no additional contraception; if started at the time of copper IUD removal, advise abstinence/barrier contraception for 7d prior to removal and for 2d afterwards
- **After childbirth** Start any time. Does not affect lactation. No additional contraception needed if <21d after childbirth. Otherwise, start as if no previous hormonal contraception

Directions for taking the POP Take 1 tablet every day with no pill-free breaks. Take each tablet at the same time each day—if delayed >3h (>12h for desogestrel POP) treat as missed pill.

Missed pills If a pill is missed/delayed >3h (>12h for desogestrel POP), continue taking at the usual time and use additional barrier methods for 2d.

⚠ Give emergency contraception with Cu-IUD or levonorgestrel if ≥1 POPs have been missed or taken >3h late (>12h late for desogestrel POP) and unprotected sexual intercourse has occurred in the 2d following this.

Diarrhoea/vomiting Continue taking the POP but use an additional barrier method during the episode and for 2d afterwards.

Interactions with other drugs Efficacy of POPs is not affected by antibacterials that do not induce liver enzymes. Efficacy is ↓ by enzyme-inducing drugs (➔ p. 734)—advise women to use an additional barrier method or alternative contraceptive method during treatment and for >4wk afterwards. Advise an alternative method of contraception if taking long-term hepatic enzyme-inducing drugs.

Follow-up Review 3mo after starting the POP or changing from CHC—earlier if complications. Once established, review every 6–12mo—assess risk factors and side effects; give health education, e.g. smoking cessation advice, information about STIs, information about long-acting reversible contraception; check BP.

Older women POP can be used until age of 55y when natural loss of fertility can be assumed. Alternatively, if aged >50y and amenorrhoeic, continue POP and check FSH; if FSH >30 IU/L, continue POP or a barrier method for 1y and then stop.

Injectable progestogens Useful if oestrogen-containing preparations are contraindicated or poor compliance. Failure rate is <4/1000 women over 2y.

Advantages

- Can be used to age 50y if no other risk factors for osteoporosis
- ↓ ectopic pregnancy, functional ovarian cysts, and sickle cell crises
- ↓ risk of endometrial cancer
- May alleviate premenstrual syndrome and ↓ menorrhagia

Disadvantages

- Relatively contraindicated if DM with complications or multiple risk factors for CVD

- May ↓ bone density in first 2–3y of use. Consider DEXA scan in older women if result would influence choice
- May be a delay in return of fertility of up to 1y on stopping
- Can cause menstrual disturbance—if troublesome give next injection early (8–11wk after the previous injection for DMPA) or add oestrogen if no contraindications
- Other side effects, e.g. weight ↑ (up to 2–3kg), mood swings, acne

Preparations

- Depo-medroxyprogesterone acetate (DMPA) 2 formulations:
  - Depo-Provera® (150mg/1mL) for deep IM administration into the buttock/lateral thigh or deltoid (do not rub the injection site afterwards), and
  - Sayana Press® (104mg/0.65mL) for sc administration into the lower abdomen or anterior thigh
- Norethisterone enantate Noristerat® (200mg/1 mL) for deep IM injection into the gluteal muscle Do not rub the injection site afterwards

⚠ *Risk of osteoporosis with medroxyprogesterone* In all women, weigh benefits of use for >2y against risks:

- If ↑ risk of osteoporosis, consider another method of contraception
- In adolescents, medroxyprogesterone should only be used when other methods of contraception are unacceptable/inappropriate for the woman

Starting injectable progestogens

- DMPA Ideally, give the first injection up to day 5 of the woman's cycle. If given for the first time after day 5 of the cycle, check the woman is not pregnant, give the first injection, and advise the woman to use an additional form of contraception for 7d (14d if 'quick starting' after UPA emergency contraception). Postpartum, give the first injection <21d after delivery if possible; check the woman is not pregnant and use additional contraception for 7d if ≥21d postpartum (if breastfeeding and amenorrhoeic, no additional contraception is needed)—➔ p. 816
- Norethisterone Give up to day 5 of the cycle or immediately after childbirth (avoid breastfeeding if baby has jaundice requiring treatment)

Repeat injections

- Depo-Provera® Every 12wk (±5d). 💣 FSRH recommends injection interval of 13wk ± 7d
- Sayana Press® Every 13wk (± 7d). Women may be taught to self-administer Sayana Press® themselves. A demonstration video and a free text reminder service are available from the manufacturer
- Noristerat® May be repeated once only after 8wk (±14d). Unlicensed if repeated further

Missed injection and no risk of pregnancy[G] ≤14wk since last injection of DMPA (≤10wk if norethisterone) or no UPSI since injection was due (i.e. no sex or barrier method used). No need for emergency contraception. Give injection immediately. If >14wk since last injection of DMPA (>10wk if norethisterone)—advise no sex or additional barrier contraception for 7d after injection

Missed injection and risk of pregnancy^G UPSI >14wk after last injection of DMPA was given (>10wk if norethisterone):

- If UPSI and last injection ≤14wk + 5d ago (≤10wk + 5d for norethisterone) or UPSI in the past 5d, give emergency contraception with Cu-IUD or levonorgestrel (➔ p. 726)
- If last injection >14wk + 5d ago (>10wk +5d for norethisterone) and UPSI >5d ago, do not give emergency contraception
- Check high-sensitivity urine pregnancy test ≥21d after last UPSI
- Consider quick start bridging contraception (e.g. POP, CHC) until −ve pregnancy test (➔ p. 728)
- Give injection once pregnancy is excluded; advise no sex or additional barrier for 7d after injection (unless Cu-IUD used as emergency contraception when leave *in situ* until 7d after injection)

Interactions Effectiveness of Noristerat® (but not DMPA) is ↓ by enzyme-inducing drugs—advise additional contraception while taking these drugs and for 4wk after stopping or alternative method.

Progestogen implant Nexplanon® is the only implant currently available in the UK. It is a radio-opaque flexible rod (40mm × 2mm) containing 68mg of etonogestrel that is inserted subdermally into the lower surface of the upper arm on day 1–5 of the cycle. If inserted after day 5, check not pregnant and use an additional method for 7d.

Advantages

- Lasts 3y and, once inserted, no compliance required
- Can be used for women at risk of ectopic pregnancy
- No effect on bone density
- Once removed, fertility returns immediately to normal

Disadvantages

- Special training is needed to insert/remove implants. Complications of minor surgery can occur (e.g. infection, scarring)
- ↓ efficacy with liver enzyme-inducing drugs—advise additional method of contraception for the duration of treatment and 4wk afterwards or alternative contraception if enzyme-inducing drugs are being used long term
- Cannot be used as part of a HRT regimen
- May cause menstrual disturbances—exclude other causes. Treat with oestrogen, additional progestogen, or NSAID
- Other side effects include acne, mood swings, breast tenderness, change in libido—treat symptoms as needed

Further information

BNF Treatment summary: contraceptives, hormonal. 🖱 https://bnf.nice.org.uk/treatment-summary/contraceptives-hormonal.html

FSRH (2016) UK MEC 🖱 www.fsrh.org/ukmec

FSRH (2014) Progestogen-only injectable contraception. 🖱 www.fsrh.org/standards-and-guidance/documents/cec-ceu-guidance-injectables-dec-2014

FSRH (2014) Progestogen-only implants. 🖱 www.fsrh.org/standards-and-guidance/documents/cec-ceu-guidance-implants-feb-2014

FSRH (2015) Progestogen-only pills. 🖱 www.fsrh.org/standards-and-guidance/documents/cec-ceu-guidance-pop-mar-2015

Intrauterine contraceptive devices

There are 2 types of intrauterine contraceptive device (IUCD), those containing copper and those containing progestogen.

Copper intrauterine device (Cu-IUD) Plastic or silver carrier wound with copper wire/bands. Acts by inhibiting fertilization, sperm penetration of the cervical mucus, and implantation. Pregnancy rate with IUCDs containing 380mm^2 copper is <20/1000 over 5y. A number of different devices are available (Table 21.5). Suitable for:

- Older parous women
- As second-line contraception in young nulliparous women, or
- For emergency contraception (➔ p. 726)

Intrauterine system (IUS) A number of levonorgestrel-releasing intrauterine systems are now available (Table 21.6): all release progestogen directly into the uterine cavity and act by preventing endometrial proliferation, thickening of cervical mucus, and suppression of ovulation (some women and some cycles). They are licensed for use in women aged >18y for

- Contraception
- Menorrhagia (menstrual bleeding is ↓ significantly in 3–6mo—➔ p. 684)
- Prevention of endometrial hyperplasia with oestrogen therapy

Contraindications

Cu-IUD Allergy to copper; Wilson's disease; heavy/painful periods.

Table 21.5 Copper intrauterine devices available in the UK

Name of device	Time to replacement (y)	Uterine length
GyneFix®	5	All
Mini TT380® Slimline	5	≥5cm
Flexi-T® 300	5	>5cm
Flexi-T® +380	5	>6cm
Multi-Safe® 375	5	6–9cm
Multiload® Cu375	5	6–9cm
Neo-Safe® T380	5	6.5–9cm
Nova-T® 380	5	6.5–9cm
Ancora® 375 Cu	5	>6.5cm
Load® 375	5	>7cm
Novaplus T 380® Cu	5	*Mini*: ≥5cm *Normal*: 6.5–9cm
Novaplus T 380® Ag	5	*Mini*: ≥5cm *Normal*: 6.5–9cm
UT380®	5	*Short*: 5–7cm *Standard: 6.5–9cm*
T-Safe® 380A QL	10y	6.5–9cm
Copper T380A®	10y	6.5–9cm

Table 21.6 Progestogen-releasing intrauterine systems

IUS	Dose of levonorgestrel	Effective for (y)	Licensed indications
Kyleena®	19.5mg (17.5 micrograms/24h)	5	Contraception
Jaydess®	13.5mg (14 micrograms/24h)	3	Contraception
Levosert®	52mg (20 micrograms/24h)	4	Contraception Menorrhagia
Mirena®	52mg (20 micrograms/24h)	5	Contraception Menorrhagia
		4💣	Endometrial protection

💣 Licensed for 4y for endometrial protection; FSRH states can be used for 5y.

Cu-IUD and IUS

- Pregnancy or <4wk postpartum
- Current or high risk of STI or pelvic inflammatory disease (includes severe immunosuppression)—do not fit an IUCD <3mo after treatment of a pelvic infection. Following treatment of STI suitability depends on ongoing risk
- Undiagnosed uterine bleeding or distorted uterine cavity
- Endometrial, ovarian, or cervical cancer, or trophoblastic disease
- Anticoagulation—caution—care needed when fitting

Advantages

Cu-IUD

- No systemic side effects and does not mask the menopause
- If fitted as emergency contraception, can provide ongoing contraception
- For women >40y, can remain in the uterus until menopause (unlicensed)

IUS

- ↓ menorrhagia/dysmenorrhoea
- ↓ risk of pelvic inflammatory disease—particularly younger age groups
- ↓ risk of ectopic pregnancy compared to the Cu-IUD
- If 45y and amenorrhoeic, can be left *in situ* for 7y for contraception (unlicensed)—change after 4y if using IUS for endometrial protection

Cu-IUD and IUS

- Long-lasting and can be used until the menopause; once fitted, no compliance is needed and easily/immediately reversible by removal
- Can be used for women who are breastfeeding, obese, or have concurrent illness—migraine, venous thromboembolism, DM, cardiovascular disease, or taking long-term enzyme-inducing drugs (e.g. anticonvulsants, antivirals)

Disadvantages and problems

Cu-IUD

- **Ectopic pregnancy** Risk (0.02/100 women/y) is higher than if using a hormonal contraceptive method. If pregnancy occurs, there is a 1 in 20 risk of ectopic pregnancy—consider ectopic pregnancy in any woman who has a Cu-IUD and develops abdominal pain

- ↑ dysmenorrhoea/menorrhagia Most common reason for discontinuation. Exclude infection and malposition. Exclude other gynaecological causes. Treat with NSAID or tranexamic acid or consider changing to the IUS

IUS Not suitable for emergency contraception. May cause:

- Changes in pattern/duration of menstrual bleeding (spotting/prolonged bleeding) are common—warn women prior to insertion. Bleeding usually becomes light/absent within 3–6mo of insertion
- Mastalgia, mood changes, change in libido—usually resolve in <6mo
- Ovarian cysts—usually resolve spontaneously; monitor with USS

Cu-IUD and IUS

- Fitting/removal Requires training; can cause discomfort for the woman
- Expulsion/malposition Risk of expulsion is ~1 in 20. Usually occurs <3mo after insertion—Figure 21.3
- Perforation of the uterus Risk ~1 in 1000. Risk is ↑ if breastfeeding
- Pelvic inflammatory disease ↑ risk of infection <21d after insertion. Related to existing carriage of STIs. ❗ It is good practice to screen for STIs (especially chlamydia) and treat infection prior to or at the time of insertion
- Actinomyces-like organisms (ALOs) on cervical smear. Assess to exclude pelvic infection. If no signs of pelvic infection, offer choice to leave device or change it. If symptomatic, discuss antibiotic treatment with microbiology and refer to SH clinic/gynaecology for further management
- Intrauterine pregnancy Confirm with USS. Remove device at <12wk gestation whether or not the woman intends to continue the pregnancy. If pregnancy is >12wk or no threads are visible, refer to obstetrics/gynaecology

Insertion Special training is needed. The FSRH runs a training scheme (☎ 020 7724 5534; email: info@fsrh.org; 🖱 www.fsrh.org). Accreditation must be updated every 5y. First device may be inserted:

- Without additional contraceptive cover if <7d after onset of menstruation (tail end of a period is the best time; heaviest days of a period are best avoided), or immediately after miscarriage/TOP
- If not in the first 7d of the cycle—exclude pregnancy (➔ p. 726) before insertion and, for IUS (not Cu-IUD), advise additional method for 7d
- Immediately after childbirth; else fit >4wk postpartum (unlicensed <6wk)—exclusion of pregnancy and need for additional contraception depends on whether breastfeeding. If breastfeeding and amenorrhoeic, no additional contraception is needed

❗ Replacement devices can be inserted at any time.

⚠ *Uterine perforation* Usually occurs during insertion; may present later. If suspected, refer for urgent pelvic USS. Advise women to contact a doctor if any of the following symptoms/signs <3wk after insertion:

- Severe pelvic pain after insertion (worse than period cramps), persistent pain (over weeks) or pain during intercourse
- ↑ bleeding or sudden changes in periods
- Unable to feel threads (but may be palpable if partial perforation)

⚠ *Women with epilepsy* ↑ risk of seizure at the time of cervical dilation—ensure emergency drugs are available.

⚠ *Cervical shock* Rare complication of IUCD insertion. Pallor, sweating, and bradycardia. Immediately tip the woman head down with legs raised. If symptoms/bradycardia persist, give 0.6mg atropine IV.

Follow-up Perform a routine check 6wk after insertion and annually thereafter^{G}. In addition, advise to return if the woman or her partner has any discomfort, or she is concerned about vaginal bleeding or discharge.

Removal

- **Pre-menopause** If pregnancy is desired, remove at any time. Otherwise, remove after establishing a hormonal method or use barrier method or abstinence for ≥7d prior to removal. If urgent removal, provide emergency contraception if mid cycle and intercourse ≤7d previously (➔ p. 726)
- **At the menopause** Remove ≥1y after periods stop if aged >50y or after 2y amenorrhoea if aged <50y. If difficulty removing the device, try again after a 5d course of topical oestrogen (e.g. Premarin® 1.25mg od po)

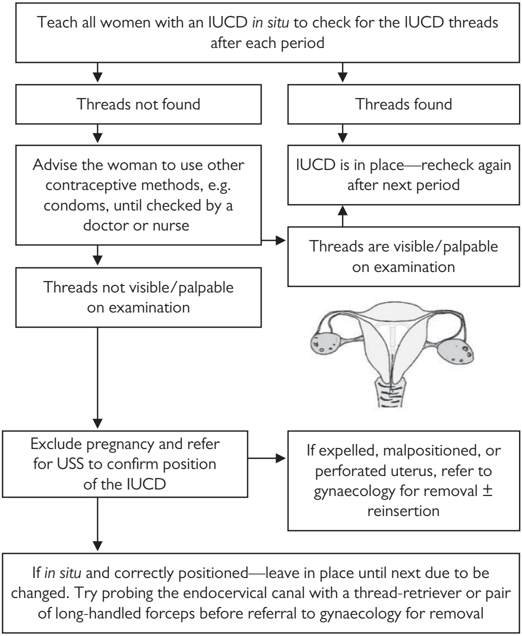

Teach all women with an IUCD *in situ* to check for the IUCD threads after each period
Threads not found
Threads found
Advise the woman to use other contraceptive methods, e.g. condoms, until checked by a doctor or nurse
IUCD is in place—recheck again after next period
Threads are visible/palpable on examination
Threads not visible/palpable on examination
Exclude pregnancy and refer for USS to confirm position of the IUCD
If expelled, malpositioned, or perforated uterus, refer to gynaecology for removal ± reinsertion
If *in situ* and correctly positioned—leave in place until next due to be changed. Try probing the endocervical canal with a thread-retriever or pair of long-handled forceps before referral to gynaecology for removal

Figure 21.3 Missing intra-uterine device threads

Reproduced from Sadler C, *et al.*, *Women's Health* (2007), with permission from Oxford University Press.

Further information

FSRH (2015) Intrauterine contraception. www.fsrh.org/standards-and-guidance/documents/ceuguidanceintrauterinecontraception
NICE (2005, updated 2019) Long-acting reversible contraception. www.nice.org.uk/guidance/cg30

Other contraceptive methods

Sterilization There are no absolute contraindications to sterilization of men or women, provided that:

- They make the request themselves
- They are of sound mind, *and*
- They are not acting under external duress

⚠ If there is any question of a person not having the mental capacity to consent to a procedure that will permanently remove their fertility, seek advice from your medical defence organization.

Method

- **Women** Laparoscopic tubal occlusion with clips or rings (usually done under GA as a day case) or hysteroscopic sterilization with intratubal implants (usually done under LA as a day case)
- **Men** Vasectomy. Usually done under LA as a day case

Pre-referral counselling

- Alternative long-term contraceptive methods (include sterilization of partner as an alternative)
- Reversibility—sterilization is intended to be permanent; reversal is only 50–60% successful
- Failure rate—1 in 200 for ♀; 1 in 2000 for ♂
- ↑ risk of ectopic pregnancy after tubal occlusion (♀)
- Risk of operative complications
- Effect on long-term health—no proven long-term risks

❗ All counselling should be supported by impartial written information.

⚠ Take additional care when counselling people:

- Aged <30y
- Without children
- Taking decisions during pregnancy
- Taking decisions in reaction to a loss of relationship
- At risk of coercion by their partner, family, or health/social welfare professionals

Need for other contraception before and after sterilization

- **Women** Other contraception until first post-procedure period
- **Men** Other contraception until semen analyses ≥8wk after the procedure shows azoospermia

Barrier methods

Condoms Give protection against STIs. Male and female versions.

- A new condom should be applied for each episode of sexual intercourse (or if applied incorrectly) and only one should be used at a time; male and female condoms should not be used simultaneously
- Advise about emergency contraception in the event of an accident
- Some lubricants/topical vaginal preparations ↓ effectiveness, e.g. petroleum jelly (Vaseline®), baby oil, and oil-based vaginal/rectal preparations; water-based lubricants are safe (e.g. KY Jelly®)

Vaginal diaphragms Latex or silicone and flat metal spring, coiled metal rim, or arcing spring diaphragms are available. Motivation is crucial. Now rarely used in the UK. Fitting must be performed by a doctor or nurse trained to fit diaphragms. If there is a weight change of >4kg, the woman has a baby, or after pelvic surgery, refer for re-fitting. Prescribe a new diaphragm yearly.

> ❗ Diaphragms should always be used in conjunction with spermicide (2 × 2cm strips applied to the upper surface)—spermicide must be reapplied if the diaphragm is *in situ* for >3h before sexual intercourse. The diaphragm must be left *in situ* for ≥6h (maximum 30h) after intercourse. Some vegetable/mineral oil-based lubricants (e.g. petroleum jelly (Vaseline®), baby oil) can damage diaphragms. Water-based lubricants are safe (e.g. KY Jelly®).

Cervical caps Silicone. Attach by suction. Otherwise used in the same way as a diaphragm. The inside of the cap should be filled one-third full of spermicide. Useful for women with poor muscle tone, absent retropubic ledge, or recurrent cystitis when using a diaphragm.

Dams Are not a contraceptive but protect against STIs. Thin film that provides a barrier between mouth/cervico-vaginal secretions or mouth/anus.

Spermicides to use in combination with diaphragms, caps, or condoms Nonoxinol '9' 2% gel (Gygel®) is the only preparation currently available in the UK.

Natural methods ⚠ Can be effective if used correctly, but failure rates are relatively high.

Coitus interruptus Penis is withdrawn prior to ejaculation.

Avoidance of intercourse during times of fertility 3 methods of estimating time of ovulation are used:

- Urine testing A commercial kit (Persona®) is available to buy
- Temperature Taken orally in the morning before drinking/getting up (thermometer is available on NHS prescription). ↑ of 0.2–0.4°C indicates progesterone release from the corpus luteum. Unprotected intercourse can take place from day 3 of the ↑ until the next period
- Mucus texture (Billing's method) Texture of vaginal secretions is felt between finger and thumb daily. Prior to ovulation the mucus becomes profuse and slippery, then abruptly changes to being thicker and more tacky. No unprotected intercourse from the day the mucus becomes more profuse until 3d after it becomes tacky. Patients with cycles > or <28d must vary timings

Further information

FSRH (2014) Male and female sterilization. 🖱 www.fsrh.org/standards-and-guidance/documents/cec-ceu-guidance-sterilisation-cpd-sep-2014
FSRH (2015) Barrier methods for contraception and STI prevention. 🖱 www.fsrh.org/standards-and-guidance/documents/ceuguidancebarriermethodscontraceptionsdi
FSRH (2015) Fertility awareness methods. 🖱 www.fsrh.org/standards-and-guidance/documents/ceuguidancefertilityawarenessmethods

Teenagers and women over 40

Contraception for the under 16s

Sexual health problems One in three have sexual intercourse under the age of 16y. Those who have intercourse early are at ↑ risk of early pregnancy and STI. Worries about sexuality can add to the pressure for some. Sensitive support, clear guidance, and accurate information about contraception, sexuality, and STIs are helpful. Remember to offer chlamydia screening (➔ p. 716) to the under 25s.

Safeguarding ➔ p. 902. ⚠ Sexual intercourse with a child under the age of 13y is rape and must be reported to the authorities.

Providing contraception to the under 16s In the UK, a doctor is allowed to give contraceptive advice and treatment to a girl aged <16y without parental consent if it is in her best interest that contraceptive advice/treatment is given and she:

- Is mature enough to understand moral, social, and emotional implications
- Cannot be persuaded to inform her parents
- Is likely to begin/continue intercourse with or without contraception
- Is likely to suffer if no contraceptive advice or treatment is given

Confidentiality and consent ➔ p. 46 and ➔ p. 49

Choice of contraceptive method

- **Condoms** Most commonly used contraception for adolescents. Relatively high failure rate—suggest their use in addition to another form of contraception to help prevent STIs
- **Long-acting reversible contraception (LARC)** Offer IUCD, progestogen implant, injectables, or intrauterine system to all teenagers. Provides high levels of protection against pregnancy with no need for ongoing compliance once fitted/administered. ❗ Medroxyprogesterone acetate preparations (Depo-Provera® and Sayana Press®) should only be used when other methods of contraception are inappropriate/unacceptable as they may ↑ osteoporosis risk (use alternative if other risk factors and try not to use >2y), and cause menstrual irregularity, and ↑ weight
- **Combined hormonal contraception (pill, patch, or vaginal ring)** Suitable method of contraception for the under 16s. Poor compliance can be a problem and leads to a relatively high failure rate
- **Progestogen-only pill (POP)** Suitable for teenagers but has the same compliance problems as combined hormonal contraception and is associated with menstrual irregularity. Useful if the teenager does not want long-acting contraception and CHC is contraindicated
- **'Morning after pill'** (levonorgestrel 1.5mg <72h or ulipristal acetate 30mg <120h after unprotected intercourse). Not suitable as regular contraception, but valuable in preventing unwanted pregnancy. Provide information on availability and make it easy for teenagers to get urgent same-day appointments to obtain emergency contraception (➔ p. 726)

Do's and don'ts

- *Don't* insist on vaginal examination unless it is necessary
- *Do* discuss the merits of delaying sexual intercourse until older

- *Do* stress the need for protection against sexually transmitted infection
- If prescribing combined hormonal contraception for acne, dysmenorrhoea, or cycle control, *do* explain its use for contraception too
- *Do* consider 'quick-starting' contraception (➲ p. 728) when the young person is seen rather than waiting for the next cycle

Information and support for teenagers

Brook Advisory Service 🖱 www.brook.org.uk
Sexwise 🖱 https://sexwise.fpa.org.uk/
Teenage Health Freak 🖱 www.teenagehealthfreak.org

Contraception for women >40y

Choice of contraceptive method

- Combined hormonal contraception (CHC) Non-smokers with no risk factors for CVD/breast cancer can use CHC until 50y; consider a lower oestrogen preparation (20 micrograms ethinylestradiol). Improves menstrual and menopausal symptoms and protects bone density. Women experiencing menopausal symptoms may prefer an extended regimen (➲ p. 732)
- Progestogen-only pill Can be continued to 55y. Does not interfere with FSH levels. Cannot be used as the progestogen component of HRT; if using as contraception, add in combined HRT and use alongside
- Injectable progestogen Can be used up to 50y in women not at risk of osteoporosis (benefits of using injectable medroxyprogesterone acetate for >2y should be evaluated against risks of ↓ bone density). May cause menstrual irregularity
- Progestogen implant Used up to 55y; do not use as part of HRT regimen
- IUS Improves menorrhagia. Licensed for endometrial protection (can be used as part of an HRT regimen). If ≥45y and amenorrhoeic, can be left *in situ* until not required for contraception (unlicensed); change after 4y (💣 FSRH states 5y) if using IUS for endometrial protection
- IUCD Copper intrauterine devices fitted in women >40y may remain in the uterus until after the menopause

Stopping contraception after the menopause

- Non-hormonal methods After 2y amenorrhoea if <50y; after 1y amenorrhoea if ≥50y
- Combined hormonal or injectable progestogen Use until 50y if no contraindications, then switch to alternative method of contraception
- Implant, POP, or IUS Continue to 50y; if ≥50y and amenorrhoeic, continue to 55y or check FSH—if FSH >30IU/L, stop contraception after 1y of amenorrhoea; if FSH is ≤30IU/L continue contraception and repeat FSH test after 1y. If 50y and not amenorrhoeic, continue contraception until 55y and then stop

Further information

FSRH (2010, updated 2019) Contraceptive choices for young people. 🖱 www.fsrh.org/standards-and-guidance/documents/cec-ceu-guidance-young-people-mar-2010

FSRH (2017) Contraception for women aged over 40 years. 🖱 www.fsrh.org/standards-and-guidance/documents/fsrh-guidance-contraception-for-women-aged-over-40-years-2017

Termination of pregnancy

The role of the GP

- The earlier in pregnancy a termination of pregnancy is performed, the lower the risk of complications. General practice is often the first stage of the referral procedure—have arrangements which minimize delay
- Termination of pregnancy (TOP), especially for 'social' reasons, is a difficult ethical area for many GPs. Whatever your views, be sympathetic, and if not prepared to refer yourself, do not pass opinion and arrange for the patient to see someone who will refer without delay
- Counselling—unbiased counselling to allow a woman to reach a decision she feels is right for her—this is an important decision she will have to live with for the rest of her life. Why does she want a termination? Has she considered alternatives? Does her partner/do her parents know? What are their views?
- Ideally the woman should be given some time once she has all the information to make her decision (e.g. follow-up in a few days). Offer a let-out clause—she can always change her mind right up until the time of the procedure, and you will support her whatever decision she makes
- Consider signing form HSA1 (remember to include your qualifications)
- Discuss contraception after TOP (ideally do this before TOP so it can be started immediately after)
- Arrange follow-up after the procedure

Legal constraints The 1967 and 1990 Human Fertilization/Embryology Acts govern termination of pregnancy in the UK. Termination is allowed at <24wk gestation if termination:

- ↓ risk to the woman's life
- ↓ risk to the mother's physical/mental health (90% TOPs are carried out under this clause)
- ↓ risk to the physical/mental health of the mother's existing children
- The baby is at serious risk of being physically or mentally handicapped

There is no upper time limit if there is:

- Real risk to the mother's life
- Risk of grave, permanent injury to the mother's physical or mental health, or
- The baby would be born seriously physically or mentally handicapped

>99% of TOPs take place <20wk. Those taking place >20wk are usually performed when fetal abnormality is found on USS or amniocentesis, or if pregnancy is concealed in the very young.

! Seek medicolegal advice from your medical indemnity organization and consider safeguarding issues if the patient is <16y or has a cognitive deficit that might impair ability to consent to referral and/or treatment.

Procedure

- Medical Oral mifepristone followed by oral and/or vaginal prostaglandin (usually misoprostol)
- Surgical Suction termination <15wk; dilatation and evacuation >15wk

Complications

- Infection
- Haemorrhage
- Uterine perforation
- Cervical trauma
- Failed procedure and ongoing pregnancy
- Psychological sequelae

❗ There is no association between TOP and subsequent infertility, miscarriage, or preterm delivery.

Follow-up In many areas post-procedure follow-up is undertaken by the GP. Worrying symptoms are: excessive blood loss, pain, *and/or* high temperature. Assess, consider the possibility of infection and treat if reasonably well; admit if the patient is unwell.

❗ Check anti-D has been given if needed (➔ p. 771) and chosen method of contraception has been started.

Contraception after termination or miscarriage <24wk

- Combined pill/patch/ring, POP, progestogen injection/implant Start on the day of surgical or second part of medical termination. No additional method required. If started >5d after termination (day 1 only for estradiol valerate/dienogest pill) an additional method is required for 2d (POP), 7d (combined pill/patch/ring or progestogen injection/implant), or 9d (estradiol valerate/dienogest pill)
- Cu-IUD or IUS Insert at time of surgical or second part of a medical abortion. No additional method required. Otherwise delay insertion to 4wk post-abortion—use another method in the interim

Teenage pregnancy The UK has the highest teenage pregnancy rate in western Europe. Not all are unplanned. Pregnant teenagers need information and non-judgemental support to help them to reach a decision whether or not to continue with the pregnancy.

Further information

RCOG (2011) The care of women requesting induced abortion. 🖱 www.rcog.org.uk/globalassets/documents/guidelines/abortion-guideline_web_1.pdf

Information/support for women about unplanned pregnancy

Antenatal results and choices (ARC) Supports parents faced with termination for fetal abnormality ☎ 0845 077 2290 🖱 www.arc-uk.org
British Pregnancy Advisory Service (BPAS) ☎ 0845 730 40 30 🖱 www.bpas.org
Brook Advisory Centres (patients <25y only) ☎ 0808 802 1234 🖱 www.brook.org.uk
Public Health England Sexwise. 🖱 https://sexwise.fpa.org.uk/
Marie Stopes International ☎ 0845 300 8090 🖱 www.mariestopes.org.uk

Infertility

Failure to conceive after 1y of regular unprotected sexual intercourse in the absence of known reproductive pathology. Affects ~1 in 7 couples.

Pregnancy rates The normal rate of pregnancy in the first year is 20–25% per cycle. 84% of couples conceive after 1y of unprotected intercourse (17 in every 20 couples); 92% conceive after 2y (19 of every 20 couples); after 3y, the pregnancy rate is still ~25%/y.

Causes of infertility

- Ovulatory dysfunction ~30%
- Pelvic disease ~20%
- Male factor ~20%
- Unknown ~30%

Initial approach Most couples tend to present at about 1y. Where possible, see the couple together. This shows mutual commitment and initiates ongoing, couple-centred management.

Couple Ask about:

- **Length of time trying to conceive**
- **Frequency of and/or difficulties with sexual intercourse** e.g. psychosexual problems, physical disability—includes excessive travelling which may limit optimal coital timing and indirectly affect fertility

Women Ask about:

- **Previous pregnancies** Children, miscarriages, same/different partner?
- **Menstrual cycle** Length of cycle (normal cycle is 21–35d duration), changes in cervical mucus through the cycle, ovulatory discomfort?
- **Past gynaecological history** Cervical smears, previous pelvic surgery, STI/pelvic inflammatory disease, PCOS
- **Past medical history** Systemic or debilitating disease, e.g. thyroid dysfunction, DM, inflammatory bowel disease, anorexia nervosa
- **Drug history** Chemotherapy, phenothiazines, cannabis, NSAIDs
- **Lifestyle** Occupation (exposure to pesticides?), smoking, alcohol, excessive exercise, stress

Men Ask about:

- **Previous children** Same/different partner?
- **PMH** Mumps, other testicular disease, STI
- **Any systemic or debilitating diseases?**
- **Drug history** Sulfasalazine, nitrofurantoin, tetracycline, cimetidine, ketoconazole, colchicine, allopurinol, α-blockers, tricyclic antidepressants, MAOI, phenothiazines, propranolol, chemotherapy, anabolic steroids, cannabis, cocaine
- **Social history** Occupation (exposure to pesticides, X-rays, solvents, paints, chemicals from smelting or welding), smoking, alcohol, excess exercise, stress, social or occupational factors which might cause testicular hyperthermia

Adverse factors Age (♀ only—fertility ↓ significantly from mid 30s), BMI <19kg/m^2 (♀ only) or >29kg/m^2 (♂ and ♀), smoking (↓ fertility by about one-third), excess alcohol (♂ only), excess caffeine (>2 cups of coffee/d—♀ only).

Examination Consider pelvic/genital examination.

GP investigations Perform investigations if no pregnancy after a year of trying to conceive—sooner if aged >35y or known cause of infertility (e.g. PCOS, endometriosis, pelvic inflammatory disease).

Female

- Rubella status
- Chlamydia serology—indicator of possible tubal disease
- Mid-luteal progesterone—check on day 21 of the menstrual cycle for a woman with a 28d cycle; adjust timing if longer/shorter cycle. Can only be interpreted after the next period as aims to 'catch' the progesterone peak 7d before the next period. Normal value (>30nmol/L) signifies ovulation
- FSH/LH—check on day 1–5 of the menstrual cycle
- Consider TFTs if symptoms/signs of thyroid disease, or prolactin if galactorrhoea or any suggestion of pituitary tumour

Male Sperm problems affect ~1 in 5 couples. Semen analysis is important even if the man already has children. If the first test is abnormal, advise loose trousers and underwear and repeat after 3mo—or as soon as possible if grossly abnormal. ❗ Abnormal sperm do not fertilize ova.

Instructions for producing a semen sample for analysis No sex for 2d beforehand and 7d since last sex (may affect motility). Masturbate into labelled sterile pot without use of condoms/gels. Keep the sample warm (e.g. inside pocket), and deliver to the laboratory within 2h. Hand over directly to a member of laboratory staff if possible.

Referral Local protocols vary and exclusions may apply. Generally refer after 18mo of failure to conceive despite regular intercourse. Refer sooner if abnormal history, examination, or investigations, e.g.:

- Female Age >35y; amenorrhoea/oligomenorrhoea; PCOS; previous pelvic inflammatory disease or STI
- Male Previous genital pathology or urogenital surgery; varicocele; significant systemic illness; persistent abnormality on semen analysis

Treatments GPs may need to continue to prescribe ❗ All treatments for infertility should be initiated in a specialist clinic:

- Clomifene—ovarian stimulation, treatment for oligospermia
- Tamoxifen—may be prescribed to women intolerant of clomifene
- Metformin—used as an adjunct to clomifene in overweight ladies with polycystic ovarian syndrome who fail to respond to clomifene alone
- Others, e.g. gonadotrophins, dopamine agonists (e.g. bromocriptine)

Counselling Consider early referral for specialist counselling. Access is through national support groups and at local specialist fertility centres. Useful contacts:

- British Infertility Counselling Association 🖱 www.bica.net
- British Fertility Society 🖱 www.britishfertilitysociety.org.uk

Further information

NICE (2013, updated 2017) Fertility problems: assessment and treatment 🖱 www.nice.org.uk/guidance/cg156

Advice and information for patients

Fertilitynetwork UK ☎ 01424 732361 🖱 http://fertilitynetworkuk.org/
The Fertility Foundation 🖱 www.fertilityfoundation.org

Sexual problems

Sexual problems may have a physical or psychological basis but all develop a psychological aspect in time. Both partners have a problem in ~30% cases. Be supportive—your response will determine whether the patient receives appropriate help.

Assessment

- **History of the problem** What is the problem? If new, when did it start? Why consult now? What outcome does the patient want? Is the patient complaining or is his/her partner?
- **Sexual history** Details of sex education; attitude towards sex; past history of sexual problems (or lack of problems)
- **Medical history** Chronic disease; psychiatric problems; medication
- **Social history** And recent life events
- **Examination** Genitalia for abnormalities or tenderness—helpful but do not insist as it may scare the patient away

! *Always consider psychological aspects* Poor self-image; anger or resentment—relationship/financial difficulties, children, parents, work stress; ignorance or misunderstanding; shame, embarrassment, or guilt—view that sexuality is 'bad', sexual abuse; anxiety/fear about sex—fear of closeness, vulnerability, letting go, and failure.

Lack of sexual interest Usually needs specialist help. Often there are underlying psychological difficulties which may relate specifically to sex, e.g. previous child abuse, or a general psychological disorder. Women frequently lose interest around the menopause or after operations (especially mastectomy or hysterectomy) or if their partner's performance repeatedly leads to frustration (e.g. impotence). Both sexes lose interest if depressed or after traumatic events.

Vaginismus Usually apparent at vaginal examination—severe spasm of the vaginal muscles and adduction of thighs. May be detected incidentally when undertaking routine procedures, e.g. cervical smear. Try to find the root cause. *Common causes*:

- Fear of the unknown
- Local pain
- Past history of rape, abuse, or severe emotional trauma
- Defence mechanism against growing up

Management Treat any underlying medical disorder causing pain. Desensitize by encouraging the woman to examine herself, and also encourage the partner to be confident enough to insert a finger into the vagina. If no success, refer.

Orgasmic problems in women *Consider*:

Physical reasons

- Drugs—major tranquillizers, antidepressants
- Neurological disease
- Pelvic surgery—recognized complication of hysterectomy

Psychological reasons

- **Women who have never achieved an orgasm** May have psychological reasons. Give 'permission' for the woman to investigate her body's own responses further by masturbation or vibrator. When she has learned how to relax, encourage her to tell her partner and incorporate caressing into their usual lovemaking
- **Women who have lost the ability to achieve orgasm** May need specialist help, especially about current relationship or loss of self-image

Sexual assault and rape ➔ p. 88 **Gender dysphoria** ➔ p. 758

Erectile dysfunction ➔ p. 754 **Dyspareunia** ➔ p. 690

Priapism with alprostadil Table 21.8, ➔ p. 755

Premature ejaculation Ejaculation sooner than either partner wishes. With practice men can learn to delay ejaculation. The stop/start technique may be effective: when during caressing or intercourse, a man feels he is close to climax he should stop being stimulated and relax for 30s; stimulation can then recommence until he is close to climax again, when the relaxation is repeated. If this fails, the woman should squeeze the penis at the base of the glans between finger and thumb during relaxation phases. Consider referral for sex therapy if no improvement.

Delayed ejaculation May be a sign of long-standing sexual inhibition. Often patients can ejaculate by masturbation but not intravaginally. Explore anxiety and guilt feelings. Use a strategy like that for erectile dysfunction (➔ p. 754). If that fails, refer for psychosexual counselling.

Retrograde ejaculation Semen passes into the bladder rather than the urethra—complication of TURP or bladder neck incision. May also occur as a result of spinal injury or DM. The patient can usually achieve an orgasm but there is no ejaculate or the volume of the ejaculate is ↓. Urine may be cloudy after having sex. Confirm diagnosis with urine microscopy (excess sperm in urine). Unless infertility is a problem, no treatment is required.

Haematospermia Blood in the ejaculate. Common causes include urogenital infection and minor urethral trauma, but often no cause is found. If persistent, underlying pathology is more likely. Ask about other symptoms, e.g. discharge, pain, dysuria. Examine the external genitalia and perform DRE to assess the prostate. Check MSU and semen analysis ± urethral swab (including chlamydia) if any urethral discharge/high risk of STI. Check PSA and urine cytology if patient is aged >40y. If persists >3mo and no cause is found, refer to urology.

Further information about specialist doctors/therapists

College of Sexual and Relationship Therapists 🖱 www.cosrt.org.uk
Institute of Psychosexual Medicine 🖱 www.ipm.org.uk

Erectile dysfunction

50% men aged 40–70y experience inability to obtain/maintain sufficient rigidity of the penis to allow satisfactory sexual performance; 90% are too embarrassed to seek help—always ask. Incidence ↑ with age.

Organic causes (80%)

- **Cardiovascular** CHD ↑ incidence of erectile dysfunction (ED) ×4—more likely to have multivessel than single-vessel coronary artery disease; peripheral vascular disease; hypertension—incidence ED is ↑ ×2
- **DM** Incidence of ED ↑×3. >35% of diabetic men have erectile dysfunction. May be the presenting feature of DM
- **Neurological** e.g. pelvic surgery, spinal injury, multiple sclerosis
- **Side effects of prescription drugs** Consider changing medication if onset of erectile dysfunction is within 2–4wk of initiation of drug therapy e.g. thiazides, SSRIs
- **Smoking** (incidence of ED is ↑ ×2) Alcohol, or drug misuse
- **Peyronie's disease** (➔ p. 438)
- **Other endocrine** Testosterone deficiency or hyperprolactinaemia

Psychogenic causes

- Performance anxiety
- Depression or stress
- Relationship failure
- Fear of intimacy

Drugs causing erectile dysfunction include:

- Antihypertensives
- Antidepressants (e.g. SSRIs)
- Major tranquillizers
- Anti-androgens
- Finasteride
- Cimetidine

History Ensure the presenting problem is erectile dysfunction and not other sexual difficulties; identify risk factors and distinguish psychogenic from organic causes (Table 21.7). ❗ Many with organic erectile dysfunction develop a psychogenic component perpetuating symptoms.

Examination and investigation

- CVD and DM—check BP, peripheral pulses, and blood for fasting lipid profile and glucose
- Psychological distress—consider depression/anxiety screening
- Testosterone insufficiency—genitals (small/absent), breasts ↑, ↓ beard (↓ frequency of shaving). If suspected, check serum testosterone, sex hormone binding globulin, free androgen index, FSH/LH ± prolactin

Table 21.7 Is erectile dysfunction psychogenic or organic?

	Psychogenic origin	Organic origin
Onset sudden or gradual?	Sudden onset	Gradual onset
Consistent loss of erections?	Inconsistent response	Consistent failure
Does the patient ever wake with an erection?	Early-morning erections	Loss of early-morning erections
Does the patient want to have intercourse?	Relationship problems	Normal libido
Age?	Usually <60y	Usually >60y

Table 21.8 Treatment options for erectile dysfunction

Treatment	Notes
Oral drugs	
Phosphodiesterase type 5 (PDE5) inhibitors, e.g. sildenafil	First-line treatment. Effective for >80%. Acts by ↑ blood flow to the penis. Sexual stimulation is still required. Avanafil, sildenafil, and vardenafil are short acting and used prn before intercourse (Table 21.9). Tadalafil is longer acting and can be used prn or taken od at lower dose to allow for frequent/spontaneous sexual activity. ⚠ *Do not* give a nitrate <24h after use
💣 Yohimbine ⚠ Not recommended	Herbal remedy derived from the African *Pausinystalia johimbe* tree. Systematic reviews generally show better than placebo, but amount/quality of active ingredient is variable between preparations and can be associated with serious side effects
Local drug treatments	
Intra-urethral or intracavernosal alprostadil	Intra-urethral preparation is effective for 40% and intracavernosal preparation for 80% patients. Used prn. Requires some manual dexterity. Takes ~10min to act. Penile pain is common. ⚠ *Prolonged erection and priapism* occurs in ~1%. Advise patients to try applying an ice pack to the upper-inner thigh, alternating between left/right thighs every 2min for 10min and to seek medical help if erection ≥4h
Mechanical devices	
Vacuum device	80% effective. Penis is placed in the device and air withdrawn mechanically sucking blood into the penis. Erection is maintained using a constriction band around the base of the penis
Penile prosthesis	Last resort. Inflatable or rigid. Major complication is infection
Others	
Androgen supplements	Ineffective unless documented hypogonadism. Only use with specialist advice. Exclude prostatic cancer and significant CVD first, and check PSA and haematocrit at 3, 6, and 12mo after initiation of treatment, then annually thereafter
Psychotherapy	Effective for some. Time-consuming and expensive but may avert the need for drugs and give permanent resolution

💣 **Male menopause or andropause** From ~30y, testosterone levels ↓ by ~10% every decade. At the same time, sex hormone binding globulin (SHBG) level ↑, which ↓ the amount of bioavailable testosterone further. Andropause is associated with low bioavailable testosterone levels; ~30% of men in their 50s develop symptoms.

Presentation ↓ sex drive, emotional, psychological, and behavioural changes, ↓ muscle mass and muscle strength, ↑ upper and central body fat, osteoporosis and back pain, ↑ cardiovascular risk.

Management If suspected, check total testosterone and SHBG ± FSH/LH and prolactin. If hypogonadism is confirmed, refer for specialist management with testosterone replacement.

GP management Counsel the couple about the problem, its possible causes and management (Table 21.8, ➔ p. 755, and Figure 21.4); provide written information. Consider stopping/changing regular medication that may be contributing to erectile dysfunction.

⚠ All men aged >25y with erectile dysfunction should be screened for DM, cardiac risk factors, and signs/symptoms of vascular disease.

Advice on lifestyle ↓ smoking, ↓ alcohol, weight ↓ (if obese), and ↑ exercise all improve both sexual function and cardiovascular health.

Phosphodiesterase type 5 (PDE5) inhibitors Table 21.9

- First-line treatment in primary care—titrate dose to effect (most people with DM need the maximum dose)
- Contraindicated if recent history of stroke/MI, systolic BP <90mmHg, hereditary degenerative retinal disease, ischaemic optic atrophy, or medication includes a nitrate or nicorandil (both can cause profound hypotension)
- Maximum 1 dose/day
- Side effects include headache, flushing, and acid reflux
- Try maximum dose on 6 separate occasions, with sexual stimulation, before judging non-response; refer men who fail to respond to maximum dose of ≥2 different PDE5 inhibitors for specialist assessment and advice

Referral Options:

- Specialist multidisciplinary erectile dysfunction clinic If treatment in primary care fails or is contraindicated
- Urology If physical urological abnormality resulting in erectile dysfunction (e.g. severe Peyronie's disease)
- Endocrinology Hormone abnormalities (e.g. ↓ androgen, ↑ prolactin)—treatment does not always restore potency
- Psychosexual counsellor Age <40y and no evidence of organic cause; psychosexual problem

❗ PDE5 inhibitors and NHS prescriptions

Generic sildenafil In the UK, generic sildenafil is available via NHS prescription for any man with erectile dysfunction. No prescription endorsement is needed.

Other drugs for erectile dysfunction (or branded sildenafil) are only available via NHS prescription for men:

- Already being prescribed treatment on 14.9.1998
- With any of the following conditions: prostate cancer; kidney failure; spinal cord injury; DM; MS; spina bifida; Parkinson's disease; polio; severe pelvic injury; or who have had radical pelvic surgery or a prostatectomy
- Through specialist services if severe distress due to erectile dysfunction

Endorse prescriptions with the letters 'SLS'.

Quantities The Department of Health (HSC 1999/148) recommends prescribing one treatment per week. If prescribing more, clearly record the reasons for prescribing a larger quantity in the patient's notes.

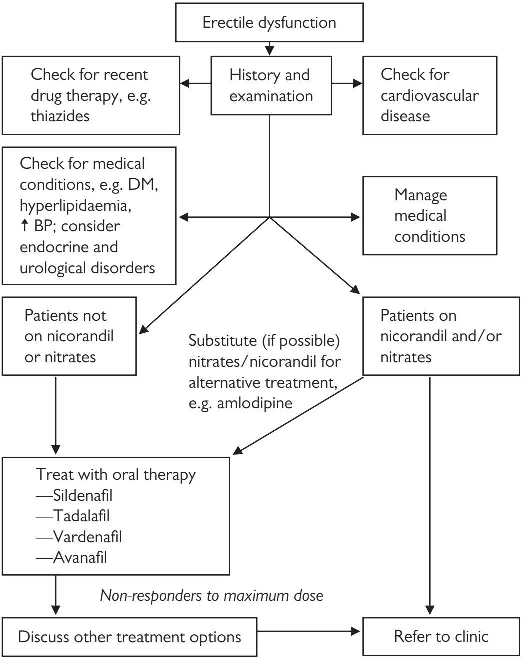

Erectile dysfunction
History and examination
Check for recent drug therapy, e.g. thiazides
Check for cardiovascular disease
Check for medical conditions, e.g. DM, hyperlipidaemia, ↑ BP; consider endocrine and urological disorders
Manage medical conditions
Patients not on nicorandil or nitrates
Patients on nicorandil and/or nitrates
Substitute (if possible) nitrates/nicorandil for alternative treatment, e.g. amlodipine
Treat with oral therapy
—Sildenafil
—Tadalafil
—Vardenafil
—Avanafil
Non-responders to maximum dose
Discuss other treatment options
Refer to clinic

Figure 21.4 Algorithm for management of erectile dysfunction

Table 21.9 PDE5 inhibitors and action times

Drug	As required dose range	Onset of action in minutes (peak action)	Duration of action (h)
Sildenafil[a]	25–100mg	20–30 (*60*)	12
Tadalafil	10–20mg[b]	60–120 (*120*)	36–48
Vardenafil[a]	10–20mg[c]	25–60 (*60*)	12
Avanafil[a]	50–200mg	15–30 (*30–45*)	6

[a] Onset of action may be delayed by up to 60min if taken with food.

[b] Dosage for regular daily dosing is 2.5–5mg od.

[c] Maximum dose 10mg if using orodispersible tablet.

Further information

BNF Treatment summary: erectile dysfunction. https://bnf.nice.org.uk/treatment-summary/erectile-dysfunction.html

Gender dysphoria and reassignment

Gender non-conformity Gender identity differs from expected cultural norms. Becomes gender dysphoria only when non-conformity causes distress.

Gender dysphoria Discomfort/distress caused by discrepancy between gender identity (psychological sense of being a man or woman) and sex assigned at birth. *Not* a psychiatric diagnosis but may cause anxiety/depression.

Transsexualism Persistent (≥2y) desire to transition and be accepted as a member of a sex other than that assigned at birth. May involve endocrine treatment ± surgery—most commonly male to female ('*trans women*' or 'MtF') but can be female to male ('*trans men*' or 'FtM').

❗ Sexual orientation (e.g. heterosexuality, bisexuality, homosexuality) is distinct and independent from gender identity.

Gender services in the UK Tertiary care services. Multidisciplinary including psychological counselling, hormone treatments, speech therapy, and surgical procedures. Goal is not always gender transition; treatment aims to enable patients to achieve a stable gender identity that they are content with.

Pre-referral tests Some gender services require baseline blood tests prior to referral. Check local protocols. Tests include:

- All Serum lipids, LFTs, bone metabolism, LH, FSH, SHBG, oestradiol, testosterone/dihydrotestosterone. *In addition*:
- MtF Prolactin, PSA
- FtM FBC

Pre-referral lifestyle modification

- Smoking ↑ risk of thromboembolism with oestrogen and polycythaemia with testosterone therapy; advise/provide assistance to stop—➔ p. 156
- Alcohol Advise to ↓ consumption to ≤14 units/wk to ↓ risk of hepatotoxicity with hormone treatments
- Substance misuse Treatment will not be initiated unless stable
- Obesity Encourage weight ↓. Obesity ↑ thromboembolic and surgical risk and may be a contraindication for surgery
- Implications for fertility Explore before starting hormone treatment. Gamete storage may be an option—take specialist advice

Referral of adults (>18y) Local referral protocols vary. Refer all patients with gender dysphoria. No upper age limit. In some areas a mental health assessment and/or application for funding may be required prior to referral.

Referral of children and young people <18y Always refer via local child and adolescent mental health services.

Prescribing hormones Do not suggest self-administration of hormones before gender clinic referral; self-medication results in poorer outcomes. Consider bridging prescriptions until seen by a specialist service, but only with specialist advice unless you have specific competencies.

Shared care GPs may be asked to continue prescriptions for oestrogen/testosterone supplements ± GnRH analogues initiated in gender clinics. Follow shared-care protocols. ❗ Ensure patients are aware that drugs are unlicensed, changes may be irreversible, and of common/serious side effects.

Table 21.10 Long-term follow-up for trans patients[G]

	Male-to-female	Female-to-male
Medication review	Hormone dose Other medication	Hormone dose Other medication
Blood tests	Annual lipids, LFTs, prolactin, oestradiol, PSA (if aged >40y)	Annual lipids, LFTs, FBC, testosterone
Routine screening	Breast, AAA	Breast, cervix (if breast tissue/cervix retained)
Other checks	Annual digital rectal prostate cancer check	If uterus/ovaries still present, refer for pelvic USS every 2y

Surgery Only considered after ≥2y of living/working in preferred gender. Various procedures are available including genital surgery, throat surgery to alter voice, and cosmetic surgery to alter appearance.

Long-term follow-up (Table 21.10) It is important for the original gender of the patient to be flagged on new notes to prevent missed diagnoses, e.g. abdominal pain in a trans-man may be a gynaecological problem. ❗ Gender-specific screening invitations will not happen automatically once the patient has a new gender NHS record. Add recalls as needed.

Changing name and gender Many informally change their name/gender when considering gender transformation.

- If official name change is needed A 'change of name certificate' countersigned by a witness may suffice. An 'Enrolled Deed Poll'' registered with the Royal Court of Justice may be needed for more formal uses. The process is slightly different in Scotland. ❗ Not available to non-UK nationals
- The Gender Recognition Act (2005) To apply for a 'Gender Recognition Certificate', individuals must be aged >18y, have lived for ≥2y and intend to live in their acquired gender lifelong and have gender dysphoria

The GP's role It is difficult to know how to address trans patients—ask. Take particular care with written correspondence/telephone as others may be unaware of gender circumstances. When making referrals, do not disclose gender history unless relevant. Ensure name changes (official or unofficial) are promptly made to patient notes. To change gender on medical records, write to the PCO registrations office together with a signed patient declaration. A new NHS number is created and the patient's old record copied to the new.

Further information

Ahmed S et al. (2013) Gender dysphoria services: a guide for GPs and other health care staff. 🖱 www.mermaidsuk.org.uk/assets/media/gender-dysphoria-guide-for-gps-and-other-health-care-staff.pdf

GOV.UK Apply for a Gender Recognition Certificate. 🖱 www.gov.uk/apply-gender-recognition-certificate

GOV.UK Change your name by deed poll. 🖱 www.gov.uk/change-name-deed-poll

National Records of Scotland Recording changes of forename(s) and surname(s) in Scotland. 🖱 https://www.nrscotland.gov.uk/registration/recording-change-of-forename-and-surname-in-scotland

Chapter 22

Pregnancy

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Pre-conception and early pregnancy counselling

The aim of pre-pregnancy care is to give a woman enough information for her pregnancy to occur under the optimal possible circumstances:

Smoking ↓ ovulation, ↓ sperm count, ↓ sperm motility.

Once the woman is pregnant Smoking is associated with:

- ↑ miscarriage rate (×2) and risk of ectopic pregnancy
- ↑ risk of cleft deformities and ↓ birth weight (by an average of ~200g)
- ↑ risk of placenta praevia and placental abruption
- ↑ risk of premature rupture of membranes and preterm delivery
- ↑ perinatal mortality

Once the baby has delivered Smoking is associated with:

- ↑ rate of cot death
- ↑ chest infections and otitis media in children

27% of pregnant women are smoking at the time of delivery. Explain risks and advise on ways to stop—➔ p. 156.

Alcohol Fetal alcohol syndrome (growth restriction, CNS involvement, and facial deformity) is rare and tends to occur in babies of heavy drinkers—especially those who binge drink. Effects of smaller quantities of alcohol are less clear. Miscarriage rates are ↑ in moderate drinkers. Advise women to avoid alcohol in pregnancy completely; if a woman continues to drink despite best advice, advise her to limit consumption to <1u/d.

Illicit drugs Cannabis (used by 5% mothers) is associated with poorer motor skills in children and strongly linked with cigarette smoking—discourage. If taking other illicit drugs, refer for specialist care.

Diet Box 22.1.

Folate supplementation ↓ risk of neural tube defect (open spina bifida, anencephaly, encephalocele) by 72%. For most women, recommend 0.4mg (400 micrograms) daily from when pregnancy is being planned until 13wk gestation. Recommend 5mg daily if:

- Previous child had neural tube defect
- Maternal/paternal history or other family history of neural tube defect
- Mother has coeliac disease, DM, BMI >30kg/m^2, or is taking anticonvulsants

Only ~1 in 3 women take folic acid prior to conception. Effect of starting in early pregnancy is unevaluated. Supplements can be prescribed, or are available via the Healthy Start Scheme (together with vitamin C and D supplements), or OTC from pharmacies/supermarkets.

Other supplements

- Vitamin D Consider 10 micrograms (400IU)/d, particularly if poor diet, limited exposure to sunlight, of South Asian, African, Caribbean, or Middle Eastern family origin, and if BMI ≥30kg/m^2
- Iron supplements Do not offer routinely—for most, side effects outweigh benefits (➔ p. 800)

Box 22.1 Healthy eating tips for women

Foods to eat Eat a variety of foods including:

- Plenty of fruit and vegetables—≥5 portions per day
- Plenty of starchy foods, e.g. bread, pasta, rice, or potatoes
- Protein-rich foods, e.g. lean meat, chicken, fish, eggs, beans, lentils
- Fibre, e.g. wholegrain bread, pasta or rice, fruit, and vegetables
- Dairy foods containing calcium, e.g. milk, cheese, and yoghurt

Folic acid (folate) ↓ risk of neural tube defects. Take folic acid supplements (400 micrograms) every day from stopping contraception until 13wk pregnant. Eat foods containing folate, e.g. green vegetables, brown rice, fortified bread, and cereals.

Iron Eat iron-rich foods, e.g. red meat, beans, lentils, green vegetables, and fortified cereals. Fruit, fruit juice, and vegetables help with iron absorption.

Foods to avoid

Pâté and some unpasteurized dairy products All pâtés (including vegetable), Camembert, Brie, other ripened soft cheeses, and blue cheese may contain *Listeria* which causes miscarriage, stillbirth, and infections in neonates.

Raw/undercooked meat, eggs, and ready meals Risk of food poisoning:

- Wash hands after handling raw meat
- Keep raw meat separate from foods ready to eat
- Only eat well-cooked meat—hot right through, with no pink bits left
- Only eat eggs cooked until white and yolk are solid. Shop mayonnaise and mousses are safe but avoid home-made dishes containing raw egg
- Ensure ready meals are piping hot all the way through

Liver products and vitamin A supplements Too much vitamin A can harm a baby's development. Avoid eating liver (and liver products, e.g. pâté) and supplements containing vitamin A or fish liver oils.

Some types of fish Eat ≥2 portions of fish per week (including 1 of oily fish—mackerel, sardines, fresh—not canned—tuna, or trout) but:

- Avoid shark, swordfish, or marlin, and limit tuna to 2 steaks or 4 cans weekly. Mercury in these fish can harm a baby's nervous system
- Only eat 1–2 portions of oily fish per week
- Avoid raw shellfish, as they can cause food poisoning

Alcohol and caffeine Avoid alcohol. High caffeine levels can cause miscarriage or ↓ birth weight. There is caffeine in coffee, tea, chocolate, cola, and some 'high-energy' drinks. Advice <4 cups of coffee, <6 cups of tea, or <8 cans of caffeinated cola daily.

Gardening and changing cat litter Toxoplasmosis can harm an unborn baby's nervous system and/or cause blindness. The parasite that causes it is found in meat, cat faeces, and soil. Advise women to wear gloves when gardening or changing cat litter and to wash their hands afterwards.

Sexual intercourse and contraception Sexual intercourse is not harmful during pregnancy. Often women contemplating pregnancy are still using contraception. Discussion about how to stop/what to expect is helpful (e.g. injectables, IUCD). Pregnancy with IUCD *in situ*—➲ p. 742.

Exercise ➲ p. 778

Chronic disease Review of pre-existing medical conditions with referral for expert pre-conceptual advice where necessary.

- Diabetes mellitus—refer for specialist diabetic review and change women taking sulfonylureas to metformin or insulin (➲ p. 806)
- Epilepsy—refer for specialist review of medication (➲ p. 807). ⚠ Counsel against pregnancy and ensure adequate contraception if taking sodium valproate due to high risk of fetal malformation
- Heart disease—refer for specialist advice if situation is not clear
- Genitourinary disease (e.g. HIV, genital warts, bacterial vaginosis)—treat and/or refer as needed for advice on mode of delivery (➲ p. 796)

Review of medication Drug handling by the body is altered during pregnancy, and drugs can cause damage to the developing fetus:

- Discontinue known teratogens prior to conception
- Avoid OTC medication unless safety checked with doctor/midwife
- Avoid prescribed medication as much as possible—few medicines have proven safety in pregnancy. If prescribing, use well-known and tested drugs at the smallest possible doses, and only when benefit > risk

Problems in previous pregnancies

- Recurrent miscarriage and/or cervical incompetence (➲ p. 767)
- Problems during delivery—early specialist referral for discussion of options
- Congenital abnormalities/inherited disorders—pre-pregnancy counselling and detailed advice on genetic screening for high-risk pregnancies is available via fetal medicine units and regional genetics services

Rubella status Rubella in early pregnancy carries a high chance (40–70%) of deafness, blindness, cardiac abnormalities, or multiple fetal abnormalities (➲ p. 792). Offer a check for rubella status if unknown. If not immune, suggest MMR vaccination; avoid pregnancy for 3mo afterwards (live vaccine) and recheck immunity after 3mo.

Flu vaccination Pregnancy ↑ risk of complications and death from seasonal influenza. Offer annual flu vaccination to all pregnant women.

Work/benefits Discussion of benefits available during pregnancy (Table 22.1) and employment law (➲ p. 778) can be helpful so that women avoid possible hazards at work, attend for antenatal care, and plan their maternity leave from early in pregnancy.

Discussion of antenatal care and screening available

- Brief discussion of antenatal screening (➲ p. 782) and antenatal care procedures (➲ p. 772) allows women to investigate their choices in pregnancy at their leisure
- Brief discussion about miscarriage and possibility of infertility allows women to be more confident about asking for help if problems with conception or early pregnancy occur

Table 22.1 Benefits available to pregnant women

Benefit	Notes
Statutory Maternity Leave (SML) and Shared Parental Leave (SPL)	• Women having a baby must take 2wk leave after the birth (4wk if factory worker) • SML allows women to take up to 52wk off work when having a baby • Apart from the first 2wk after birth (which must be taken by the mother), that leave can be taken by the mother alone or shared with the child's other parent if both meet eligibility criteria (SPL) • SML starts the day after the birth, or any time from the 11th week prior to the due date • Automatically starts if off work for a pregnancy-related illness ≤4wk before the week the baby is due ❗ In addition, women are entitled to paid time off for antenatal care
Statutory Maternity Pay (SMP) and Shared Parental Pay (ShPP)	• Weekly sum paid for up to 39wk • Must have worked for the same employer for 26wk prior to the 15th week before the baby is due and earn on average at least the National Insurance (NI) lower earnings limit • The woman must inform her employer ≥28d before starting leave and submit claim form MatB1 signed by a midwife/GP • Maternity Pay Period (MPP) starts when SML begins • If taking SPL, can receive ShPP for a maximum of 37wk
Maternity Allowance (MA)	• Weekly sum paid for up to 39wk • For women employed but not eligible for SMP, self-employed, or who recently stopped working • Minimum work commitment, income, and NI contribution restrictions apply. ❗ Women who do not meet these eligibility criteria may be entitled to ↓ amount/duration of MA payments • Claim with form MA1 (online or download from 🖱 www.gov.uk). A MatB1 form signed by the woman's GP/midwife, proof of earnings, and SMP1 form (if SMP refused by employer) must accompany applications
Low-income benefits	Universal Credit or Income Support (➔ p. 104) may be available for women unable to claim SMP/MA
Paternity leave and pay	• 1 or 2 weeks leave to be taken from day of birth up to 56d after birth • Eligible if worked for the same employer for >26wk and earning more than NI lower earnings limit in the qualifying period • Weekly sum paid through employer • Must give notice to take leave ≥15wk before the expected due date • Can also have time off work for up to 2 antenatal appointments
Sure Start Maternity Grant	• One-off payment (currently £500) • For women claiming Universal Credit or equivalent low-income benefit • Claim from 11wk before the baby is due to <3mo after birth if no other children <16y (or if multiple pregnancy)
Free dentistry/prescriptions	• All mothers while pregnant and <1y after the expected date of delivery • Claimed using form FW8 supplied by GP/midwife
Healthy Start vouchers and free vitamins	Women claiming low-income benefits may be able to claim vouchers for milk/fruit/vegetables, and free vitamin supplements if >10wk pregnant. Download application form from 🖱 www.gov.uk or ☎ 0345 607 6823
Child Benefit	➔ p. 827

Bleeding in early pregnancy

Bleeding up to 14wk into pregnancy Bleeding in early pregnancy occurs in 1 in 4 pregnancies. *Causes*:

- Bleeding in normal pregnancy—largest group
- Miscarriage
- Ectopic pregnancy
- Trophoblastic disease
- Non-obstetric conditions, e.g. friable cervix, polyp, cervical neoplasia

> ⚠ Any sexually active woman presenting with abdominal pain and vaginal bleeding after an interval of amenorrhoea has an ectopic pregnancy until proven otherwise.

Assessment

- Take a history of pain and bleeding—pain preceding bleeding suggests ectopic pregnancy is more likely. Have any products of conception been passed? ❗ Clots/products can be difficult to distinguish
- Check LMP and pregnancy test result (do a test if needed)
- Check pulse (>100bpm suggests shock), BP, and temperature (?toxic)
- Abdominal examination—guarding, peritonism, and/or unilateral tenderness suggest ectopic pregnancy
- Pelvic examination is not necessary in primary care

Initial management If severe bleeding and/or pain, shocked, or toxic, admit to gynaecology as an emergency. If shocked, give 1mL Syntometrine® IM, if available, and try to gain IV access.

If clinically well, refer to the Early Pregnancy Assessment Unit (EPAU) to check site and viability of pregnancy with USS. Local referral criteria vary:

- At 5wk, gestation sac ± yolk sac is seen on scan
- At 6–7wk, a fetal pole and fetal heart beat is usually seen

Blood group and rhesus status is also checked at the EPAU (➔ p. 770).

Complications of bleeding Significant sub-chorionic haematoma is associated with ↑ risk of premature rupture of membranes and IUGR—refer early for specialist antenatal care.

> ⚠ **Rhesus-negative women** ❗ If clinical doubt, give anti-D.
>
> *Bleeding <12wk gestation* Anti-D is not required for:
>
> - Threatened miscarriage unless heavy or repeated bleeding and/or abdominal pain, or
> - Complete miscarriage where no medical or surgical uterine evacuation
>
> *Bleeding ≥12wk gestation, ectopic pregnancy, and/or medical/surgical evacuation of the uterus at any gestation* Give anti-D immunoglobulin within 72h of bleeding— whether or not the pregnancy is lost—➔ p. 770.

Bleeding in early normal pregnancy Often termed *threatened miscarriage*. If fetal heart is seen on USS then there is ~97% chance of the pregnancy continuing to progress. There is no evidence that rest or abstinence from sex improves outcome.

Miscarriage Also termed spontaneous abortion. Occurs in 1 in 5 pregnancies—80% at <12wk gestation. *Risk factors*:

- Maternal age ≥35y or paternal age ≥40y
- BMI >29kg/m^2—if >32kg/m^2, risk is ↑ by 30%
- Smoking
- Excess alcohol

Causes

- Fetal abnormality (50%)
- Multiple pregnancy
- Uterine abnormality—fibroids, polyps; congenital abnormality; cervical incompetence (➔ p. 768—late second-trimester miscarriages)
- Systemic disease—renal, autoimmune, or connective tissue disease—particularly SLE, PCOS, DM, systemic infection
- Drugs—cytotoxics, diethylstilbestrol
- Placental vascular abnormalities

Classification

- **Complete miscarriage** History of bleeding. No products of conception in the uterus. Provide psychological support
- **Incomplete miscarriage** Bleeding. Products of conception remain in the uterus but there is no fetal heart. May be managed surgically or medically (with prostaglandin analogues ± antiprogesterone priming). Alternatively, some women prefer a 'watch and wait' approach—at 3d 86% of threatened miscarriages will be complete
- **Missed (or delayed) miscarriage** No bleeding. Usually discovered when no heart beat is seen on routine antenatal scan. Treatment is medical or surgical as for threatened miscarriage. A 'watch and wait' approach is possible, but at 4wk only 66% are complete, and associated with longer bleeding

❗ There is no evidence that abstinence from pregnancy for a time after miscarriage is helpful—fertility may ↑ immediately after miscarriage.

Complications

- **Early** Perforation of the uterus, retained products of conception, infection. Treat with antibiotics if infection is suspected (e.g. co-amoxiclav 500mg tds). Re-refer/readmit if shock, pain, heavy bleeding, or bleeding is not settling
- **Later** Uterine synechiae (Asherman's syndrome), cervical incompetence, psychological sequelae

Recurrent miscarriage^{G} ≥3 miscarriages (1–2% couples). *Check*:

- **Age** Incidence of recurrent miscarriage ↑ with age (♂ and ♀)
- **How many miscarriages** Confirmed pregnancies (not just late, heavy periods)? All with the same partner? What gestation? The more miscarriages, the lower the chance of successful pregnancy
- **Infertility treatment?** 25–30% of women who miscarry
- **Past medical history** Gynaecological problems (cervical instrumentation, PCOS); systemic disease
- **Family history** Recurrent miscarriage, thrombosis/thrombophilia

Management of recurrent miscarriage Refer for further investigation; consider checking antiphospholipid antibodies prior to referral. No cause is found in 50% of those referred; they have ~70% chance of successful pregnancy. Other treatable causes include:

- **Antiphospholipid antibodies** (15%) Low-dose aspirin + LMWH from 6 to 34wk improves outcome
- **Inherited thrombophilia** (factor V Leiden, prothrombin gene mutation, or protein S deficiency) If recurrent miscarriage >10wk, treatment with LMWH ↑ live birth rate
- **Cervical incompetence** Diagnosis is usually made on the basis of ≥1 late 2nd- or early 3rd-trimester miscarriage (usually painless leaking of liquor or gradual painless dilatation of the cervix). Refer all women with past history for early obstetric review. Treatment is with cervical USS monitoring with cerclage if cervical length is <25mm (a stitch is placed high up around the cervix to keep it closed, e.g. McDonald suture. The stitch is removed at ~37wk and labour ensues rapidly if the diagnosis was correct)
- **Chromosomal abnormality in one parent** (3–5%) Refer for specialist genetic advice

Ectopic pregnancy[G] Embryo implants outside the uterine cavity—mostly in a fallopian tube. Incidence ~1 in 200 pregnancies. *Risk factors*:

- Pelvic inflammatory disease (single episode ↑ risk ×7)
- Infertility (15%)
- IUCD (14%)
- Previous ectopic (11%)
- Tubal surgery
- Age >35y
- Smoking
- Multiple partners

History

- **Abdominal pain** (97%) Unilateral or bilateral, may start before bleeding; radiates to shoulder tip; ↑ on passing urine/opening bowels
- **Amenorrhoea** (75%) Peak incidence after 7wk amenorrhoea
- **Irregular vaginal bleeding** (79%) Described as 'prune juice' but may be fresh blood; usually not heavy. May pass decidual cast

Examination Shock in 15–20%; abdominal tenderness ± rebound or guarding (71%); pelvis—enlarged uterus, adnexal mass, and/or cervical excitation.

Management Admit immediately for further investigation. Resuscitate before admission as needed. Hospital management may be expectant (watch and pregnancy resolves spontaneously), medical (methotrexate), or surgical (usually laparoscopic). Offer early USS in future pregnancies to confirm pregnancy is intrauterine.

Complications Death if undetected, infertility (pregnancy rate after ectopic pregnancy is ~66% with 10% having a further ectopic pregnancy).

Trophoblastic disease[G]

Hydatidiform mole Trophoblastic tumour containing 46 chromosomes (usually of paternal origin) and no fetal material. 8–20% become invasive and penetrate the uterus and/or metastasize to the lungs. Presents with:

- Bleeding in early pregnancy ± exaggerated symptoms of pregnancy
- Uterus is usually large for dates, and no fetal heart can be heard
- USS has a typical appearance; blood—↑↑ serum β-hCG
- Rarely symptoms of metastatic spread—haemoptysis, pleurisy

Refer urgently to gynaecology. If mole is confirmed, women are followed up by specialist centres. Invasive disease requires chemotherapy. Combined hormonal contraception is contraindicated until normal hCG values are obtained. Pregnancy is not advised until completion of the surveillance period—investigate with early USS and β-hCG as incidence of further molar pregnancy is ~1 in 80.

Partial mole Tumour of trophoblast containing 69 chromosomes, 1 maternal and 2 paternal sets, with some fetal tissue. A fetal heart may be seen on early USS but is absent by 8–9wk. Treat as for mole. Rarely becomes malignant (0.5%).

Choriocarcinoma Malignant trophoblastic tumour following molar (rarely normal) pregnancy. Presents with vaginal bleeding and/or metastases (shadows on CXR, dyspnoea, haemoptysis). Excellent prognosis after treatment with chemotherapy. No contraceptive restrictions after completion of therapy; pregnancy is possible >1y after treatment.

Placental site trophoblastic tumour (PSTT) Rare. Follows 3–4y after normal pregnancy. Presents with abnormal bleeding or amenorrhoea. Can present with distant metastases similarly to choriocarcinoma. Treatment is with hysterectomy ± chemotherapy. Prognosis is good.

Psychological effects of early loss of pregnancy Ask all women who have suffered early loss of pregnancy. Include the woman's partner if possible.

- Legitimize grief and acknowledge it—not all women grieve—adjust your approach accordingly; discuss worries/concerns
- Provide information about the condition which caused the loss; risk to future pregnancies (if <3 miscarriages, risk of further miscarriage is not significantly ↑; risk of further ectopic pregnancy is ~1 in 10); and self-help/support organizations, e.g. Miscarriage Association
- Warn of the anniversary phenomenon (sadness at the baby's due date or anniversary of the pregnancy loss) or sadness/jealousy on the birth of another's baby—provide ongoing support as needed
- If the woman already has young children, inform the health visitor

Further information

NICE (2019) Ectopic pregnancy and miscarriage: diagnosis and initial management. 🖱 https://www.nice.org.uk/guidance/ng126
RCOG (2010) Gestational trophoblastic disease. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg38
RCOG (2011, updated 2017) Recurrent miscarriage: Investigation and treatment of couples. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg17/

Patient advice and support

Ectopic Pregnancy Trust ☎ 020 7733 2653 🖱 www.ectopic.org
Hydatidiform Mole and Choriocarcinoma UK Information and Support Service 🖱 www.hmole-chorio.org.uk
Miscarriage Association ☎ 01924 200799 🖱 www.miscarriageassociation.org.uk

Haemolytic disease and rhesus isoimmunization

15% of women are Rhesus D (RhD) –ve. Development of anti-D antibodies results from fetomaternal haemorrhage (FMH) in RhD –ve women carrying a RhD +ve fetus. In later pregnancies, these antibodies cross the placenta causing rhesus haemolytic disease of the fetus which gets successively worse with each pregnancy.

All RhD –ve mothers are tested for D antibodies at booking, at 28wk, and 2-weekly thereafter. Testing is not performed once women are given anti-D prophylaxis. Anti-D titres <4u/mL (<1 in 16) are unlikely to cause serious disease. If >10u/mL, refer for specialist advice.

Effects on the fetus

- Hydrops fetalis (oedematous fetus)
- Intrauterine death

Effects on the neonate

- Jaundice
- Heart failure (oedema, ascites)
- Anaemia
- Yellow vernix
- Hepatosplenomegaly
- CNS signs

❗ All neonates with haemolytic disease must be managed by specialist paediatricians. Treatment involves UV light for jaundice ± exchange transfusion.

Immunoprophylaxis Immunoprophylaxis for RhD –ve mothers using anti-D immunoglobulin (anti-D Ig) is given IM into the deltoid muscle as soon as possible after the sensitizing event—preferably within 72h though there is evidence of benefit up to 10d. Women already sensitized should not be given anti-D Ig.

Test for the size of fetomaternal haemorrhage In the UK, blood is taken from the mother (anticoagulated sample) as soon as possible (preferably <2h) after the sensitizing event if >20wk gestation. A Kleihauer acid elution test detects fetal haemoglobin (HbF) and identifies women with large fetomaternal haemorrhage who need additional anti-D Ig.

Other causes Anti-D antibodies are the most common cause of rhesus disease. Other causes include: Rh C, E, c, e, Kell, Kidd, and Duffy. Anti-Du antibodies are relatively common but usually harmless. Follow advice of your local transfusion service about follow-up.

❗ Anti-D Ig rarely causes allergic reactions. If the woman is worried about use of blood products, an alternative approach is to check rhesus status of the father—if he is Rh –ve, then the baby is Rh –ve as well, so anti-D prophylaxis is not required.

Further information

BCSH (2014) Guideline for the use of anti-D immunoglobulin for the prevention of haemolytic disease of the fetus and newborn. 🖱 https://onlinelibrary.wiley.com/doi/abs/10.1111/tme.12091

When should anti-D be administered?

If threatened or spontaneous miscarriage

- ≥20wk—500IU + test the size of the FMH
- 12–20wk gestation—250IU
- <12wk gestation—only administer 250IU if bleeding is heavy, repeated, or there is associated abdominal pain (particularly if close to 12wk), or there has been an intervention to evacuate the uterus (e.g. ERPC)
- If bleeding continues intermittently after 12wk, give anti-D Ig 6-weekly

Following termination of pregnancy/ectopic pregnancy All non-sensitized RhD –ve women: 250IU if <20wk; 500IU if ≥20wk + test for the size of the FMH.

Following sensitizing events before delivery All non-sensitized RhD –ve women after:

- Invasive prenatal diagnosis (amniocentesis, chorionic villus sampling, fetal blood sampling) or other intrauterine procedures
- Antepartum haemorrhage
- External cephalic version of the fetus
- Closed abdominal injury (e.g. RTA)
- Intrauterine death

<20wk—250IU; ≥20wk—500IU + test size of FMH.

Routine antenatal prophylaxis

- 1–1.5% of RhD –ve women develop anti-D antibodies during pregnancy due to FMH which is small and silent—most commonly in the 3rd trimester
- Routine antenatal prophylaxis ↓ sensitization to <0.2% and is usual practice in the UK. Give irrespective of whether a woman has had prior anti-D prophylaxis earlier in the pregnancy
- Administration of 500IU anti-D Ig at 28wk (after blood has been taken for routine antibody screening) and 34wk gestation—or single dose of 1500IU at 28wk—↓ incidence of immunization after birth
- Women who have been given antenatal prophylaxis may still be sensitized by a large FMH so, following any potentially sensitizing event, additional anti-D Ig should be given and a Kleihauer test performed
- Screening for anti-D antibodies after prophylaxis is uninterpretable

Postnatal prophylaxis

- 500—1500IU (500IU in the UK) is given to every non-sensitized RhD –ve woman <72h after delivery of a RhD +ve infant
- >99% women have a FMH of <4mL at delivery—a test to detect FMH >4mL must be done so that additional anti-D Ig can be given as needed
- Risk factors for high FMH include: traumatic delivery, Caesarean section, manual removal of placenta, stillbirth and intrauterine death, abdominal trauma during the 3rd trimester, twin pregnancy (at delivery), unexplained hydrops fetalis

⚠ Doses of Rhophylac® differ—check the *BNF*/drug data sheet for correct dosing.

Antenatal care

Objectives of good obstetric care

- To provide a safe outcome for the mother and baby with the minimum of avoidable complications
- To make the birth experience as satisfying as possible
- To make optimal use of available resources

Pregnancy is a risky business for both mother and baby. Every year women die as a result of pregnancy—the most common causes being eclampsia, haemorrhage, pulmonary embolism, and infection.

Maternity Services Are provided by practices as an Additional Service, i.e. most practices are expected to provide routine antenatal care and postnatal care to mothers and babies from birth (or discharge from secondary care) until the 14th day after delivery, with the exception of intrapartum care and the neonatal check. Payment is included in the global sum. If a practice 'opts out', global sum is ↓ by 2.1%.

Intrapartum care Intrapartum care and neonatal checks can be provided to women by GPs at home or in GP maternity units as a National Enhanced Service. One payment is made for each woman who receives intrapartum care and a further payment for each neonatal check.

Definitions

- **Gravidity** Number of pregnancies a woman has had (at any stage)
- **Parity** Number of pregnancies resulting in delivery >24wk gestation (or live births <24wk)
- **Primipara, multipara** Woman who has been delivered of a child for the first time (primipara) or second or subsequent time (multipara)

Pregnancy tests Detect urinary β-hCG. +ve from 1st day of missed period (highly sensitive tests are +ve from 21d) until ~20wk gestation. Remain positive for ~5d after miscarriage/termination or fetal death.

Antenatal care The first antenatal appointment should be offered as early into pregnancy as possible. Further appointments for healthy women should be offered at 16, 28, 34, 36, 38, and, if not already delivered, at 41wk. Additionally healthy nulliparous women should be offered appointments at 25, 31, and 40wk. See Figure 22.2 ➔ p. 775. Provide additional appointments as needed for high-risk women.

First antenatal visit The primary function of this visit is to identify those women needing additional care. As there is so much information to be collected/discussed, often split across two appointments.

History

- This pregnancy—LMP, usual cycle, fertility problems, contraception, desirability of pregnancy, any problems so far
- Estimated date of delivery (EDD)—Figure 22.1
- Past pregnancies—outcome and complications of previous pregnancies
- Past/current medical history—illness (including psychiatric illness), drugs, allergies, varicose veins, abdominal/pelvic surgery (including female genital mutilation)
- Family history—↑ BP, DM, congenital/genetic abnormality, twins
- Social history—smoking, alcohol consumption, illicit drugs, support at home, work, housing, financial problems

Date of first day of last menstrual period							*Month →*					
Day ↓	Jan	Feb	Mar	Apr	May	Jun	Jul	Aug	Sep	Oct	Nov	Dec
1	8/10	8/11	6/12	6/1	5/2	8/3	7/4	8/5	8/6	8/7	8/8	7/9
2	9/10	9/11	7/12	7/1	6/2	9/3	8/4	9/5	9/6	9/7	9/8	8/9
3	10/10	10/11	8/12	8/1	7/2	10/3	9/4	10/5	10/6	10/7	10/8	9/9
4	11/10	11/11	9/12	9/1	8/2	11/3	10/4	11/5	11/6	11/7	11/8	10/9
5	12/10	12/11	10/12	10/1	9/2	12/3	11/4	12/5	12/6	12/7	12/8	11/9
6	13/10	13/11	11/12	11/1	10/2	13/3	12/4	13/5	13/6	13/7	13/8	12/9
7	14/10	14/11	12/12	12/1	11/2	14/3	13/4	14/5	14/6	14/7	14/8	13/9
8	15/10	15/11	13/12	13/1	12/2	15/3	14/4	15/5	15/6	15/7	15/8	14/9
9	16/10	16/11	14/12	14/1	13/2	16/3	15/4	16/5	16/6	16/7	16/8	15/9
10	17/10	17/11	15/12	15/1	14/2	17/3	16/4	17/5	17/6	17/7	17/8	16/9
11	18/10	18/11	16/12	16/1	15/2	18/3	17/4	18/5	18/6	18/7	18/8	17/9
12	19/10	19/11	17/12	17/1	16/2	19/3	18/4	19/5	19/6	19/7	19/8	18/9
13	20/10	20/11	18/12	18/1	17/2	20/3	19/4	20/5	20/6	20/7	20/8	19/9
14	21/10	21/11	19/12	19/1	18/2	21/3	20/4	21/5	21/6	21/7	21/8	20/9
15	22/10	22/11	20/12	20/1	19/2	22/3	21/4	22/5	22/6	22/7	22/8	21/9
16	23/10	23/11	21/12	21/1	20/2	23/3	22/4	23/5	23/6	23/7	23/8	22/9
17	24/10	24/11	22/12	22/1	21/2	24/3	23/4	24/5	24/6	24/7	24/8	23/9
18	25/10	25/11	23/12	23/1	22/2	25/3	24/4	25/5	25/6	25/7	25/8	24/9
19	26/10	26/11	24/12	24/1	23/2	26/3	25/4	26/5	26/6	26/7	26/8	25/9
20	27/10	27/11	25/12	25/1	24/2	27/3	26/4	27/5	27/6	27/7	27/8	26/9
21	28/10	28/11	26/12	26/1	25/2	28/3	27/4	28/5	28/6	28/7	28/8	27/9
22	29/10	29/11	27/12	27/1	26/2	29/3	28/4	29/5	29/6	29/7	29/8	28/9
23	30/10	30/11	28/12	28/1	27/2	30/3	29/4	30/5	30/6	30/7	30/8	29/9
24	31/10	1/12	29/12	29/1	28/2	31/3	30/4	31/5	1/7	31/7	31/8	30/9
25	1/11	2/12	30/12	30/1	1/3	1/4	1/5	1/6	2/7	1/8	1/9	1/10
26	2/11	3/12	31/12	31/1	2/3	2/4	2/5	2/6	3/7	2/8	2/9	2/10
27	3/11	4/12	1/1	1/2	3/3	3/4	3/5	3/6	4/7	3/8	3/9	3/10
28	4/11	5/12	2/1	2/2	4/3	4/4	4/5	4/6	5/7	4/8	4/9	4/10
29	5/11		3/1	3/2	5/3	5/4	5/5	5/6	6/7	5/8	5/9	5/10
30	6/11		4/1	4/2	6/3	6/4	6/5	6/6	7/7	6/8	6/9	6/10
31	7/11		5/1		7/3		7/5	7/6		7/8		7/10

Dates are given in the format day/month

! As a rough guide, EDD = date of LMP + 1 year + 7 days – 3 months

Figure 22.1 Expected date of delivery calculator

Examination

- Check weight and calculate BMI—low BMI ↑ risk of pre-eclampsia, IUGR, and preterm delivery; high BMI is associated with pre-eclampsia
- Listen to heart and lungs; check BP, and examine abdomen
- Fetal heart with a fetal hand-held Doppler or sonic aid per abdomen from 12–14wk gestation
- Fundus can be felt per abdomen from 12wk

Investigations

- Offer early USS at 10wk–13wk 6d for dating purposes and to detect multiple pregnancy. Offer routine anomaly scan at 18wk–20wk 6d gestation. If the placenta extends across the internal cervical os, arrange repeat USS at 32wk
- Check blood for Hb, blood group, Rhesus status, and red cell antibodies; syphilis/rubella serology, HBsAg/HIV with pre-test counselling; sickle test and/or Hb electrophoresis
- MSU for protein and bacteriuria
- Discuss and offer antenatal screening (Figure 22.3, ➔ p. 777)
- Inform women <25y about chlamydia screening through the National Chlamydia Screening Programme (➔ p. 796)

Education

- Health promotion (➔ p. 778)
- Employment rights (➔ p. 778)
- Travel and limitations (➔ p. 779)
- Antenatal/parent craft classes
- Benefits (➔ p. 765)
- Pelvic floor exercises (➔ p. 821)
- Free prescriptions/dental care (➔ p. 765)
- Local services (e.g. aquanatal classes, yoga for pregnancy)
- Choice of place of delivery and options available (➔ p. 780)
- Procedure for antenatal care—Figure 22.2

Information

- Offer 'Screening tests and your baby' to all pregnant women (available to download/order in a variety of languages from 🖱 www.gov.uk)
- Direct women to the NHS 'Your pregnancy and baby guide' (🖱 www.nhs.uk/conditions/pregnancy-and-baby) and/or NHS Scotland 'Ready, Steady Baby' (🖱 www.readysteadybaby.org.uk)
- Offer 'Emma's diary' to all pregnant women. Contains information and vouchers (order by e-mail: emma@emmasdiary.co.uk or ☎ 01628 566050). Also available online (🖱 www.emmasdiary.co.uk)

Discussion Worries about pregnancy or social situation—ask specifically about domestic violence.

Follow-up visits Ask about problems and untoward symptoms. Provide the neonatal bloodspot screening leaflet at ~28wk.

Routine checks

- BP
- Oedema
- Urine for protein
- Fundal height (from 24/25wk)
- Fetal heart sounds (from 12–14wk)
- Fetal lie and presentation (from 36wk)

❗ Primiparous women are aware of movements from ~20wk but multiparous women often feel movements earlier.

Laboratory checks Hb and antibodies at 28wk.

GESTATIONAL AGE

☐	Folic acid supplementation (400 micrograms/d until 13wk). Food hygiene and lifestyle advice (➔ p. 763). Information about antenatal screening tests (➔ p. 782).	First contact
☐	*Provide information on:* fetal development, nutrition, vitamin D supplementation, exercise (including pelvic floor exercises), antenatal screening, pregnancy care, antenatal care, breastfeeding, and maternity benefits. Identify women who need extra care. *Check*: BP, BMI, urine for proteinuria. Offer and arrange antenatal screening tests. Arrange early USS and fetal anomaly scan.	Booking <10wk May be 2 appointments
☐	Review, discuss, and record results of all screening tests. Investigate Hb <11g/dL—consider iron supplements. Measure BP and test urine for proteinuria.	16wk
▨	Measure SFH + BP. Test urine for protein.	25wk
☐	Measure SFH + BP. Test urine for protein. Offer repeat screening for anaemia and atypical red cell antibodies. Offer first dose anti-D if rhesus-negative. Investigate Hb <10.5g/dL—consider iron supplements.	28wk
▨	Measure SFH + BP. Test urine for protein. Review, discuss, and record results of 28wk screening tests.	31wk
☐	Measure SFH + BP. Test urine for protein. Offer second dose anti-D if rhesus negative. Give information about labour. For parous women, review, discuss, and record results of 28wk screening tests.	34wk
☐	Measure SFH + BP. Test urine for protein. Check presentation—offer external cephalic version if breech. Give information about postnatal care (including depression), breastfeeding, baby care, vitamin K prophylaxis, and neonatal screening.	36wk
☐	Measure SFH + BP. Test urine for protein.	38wk
▨	Measure SFH + BP. Test urine for protein.	40wk
☐	Measure SFH + BP. Test urine for protein. Offer membrane sweep. Offer induction after 41wk.	41wk

SFH = symphysis-fundal height (in cm) ☐ All women ▨ First pregnancy only

❗ Reassess the need for additional care of pregnant woman at each visit.

Figure 22.2 Algorithm for routine antenatal care

⚠ Symptoms suggesting serious pregnancy complications

- Abdominal pain
- Vaginal bleeding
- Clear vaginal loss
- Severe headache
- Blurred vision
- Persistent itch
- Changed/↓ fetal activity

Women who may need additional care

- Pre-existing medical conditions, e.g. ↑ BP, cardiac, renal, endocrine, autoimmune, or haematological disorders, epilepsy, severe asthma, DM, cancer, HIV, hepatitis B, substance abuse, female genital mutilation
- Factors that make the woman vulnerable, e.g. lack of social support, domestic violence
- Age ≥40y or ≤18y
- BMI ≥30kg/m^2 or <18kg/m^2
- Smoker
- Uterine surgery—e.g. Caesarean section, myomectomy, cone biopsy
- Previous pre-eclampsia, eclampsia, or HELLP
- Para ≥6 or ≥3 miscarriages
- Previous preterm birth, mid-trimester loss, stillbirth, or neonatal death
- Previous psychiatric illness or puerperal psychosis
- Previous baby with congenital abnormality
- Previous small-for-gestational age or large-for-gestational age baby or baby weighing <2.5kg or >4.5kg at birth
- Family history of genetic disorder

Interventions that are *not* part of routine antenatal care

- Repeated maternal weighing—only weigh if clinical management is likely to be influenced, e.g. concern about nutrition
- Breast or pelvic examination
- Iron supplementation
- Screening for chlamydia, CMV, HCV, group B streptococci, toxoplasmosis, or bacterial vaginosis
- Screening for preterm birth by assessment of cervical length (either by USS or vaginal examination) or using fetal fibronectin
- Formal fetal movement counting
- Antenatal electronic cardiotocography
- USS >24wk
- Umbilical or uterine artery Doppler USS

Further information

Fetal Anomaly Screening Programme 🖱 www.gov.uk/guidance/fetal-anomaly-screening-programme-overview
NICE (2008, updated 2019) Antenatal care: routine care for healthy pregnant women. 🖱 www.nice.org.uk/guidance/cg62

Information and support for pregnant women

Emma's diary 🖱 www.emmasdiary.co.uk
Mothers 35 plus 🖱 www.mothers35plus.co.uk
National Childbirth Trust (NCT) ☎ 0300 330 0700 🖱 www.nct.org.uk
Ready, Steady, Baby 🖱 www.readysteadybaby.org.uk
Which? Birth Choice 🖱 www.which.co.uk/birth-choice
Your pregnancy and baby guide 🖱 www.nhs.uk/conditions/pregnancy-and-baby

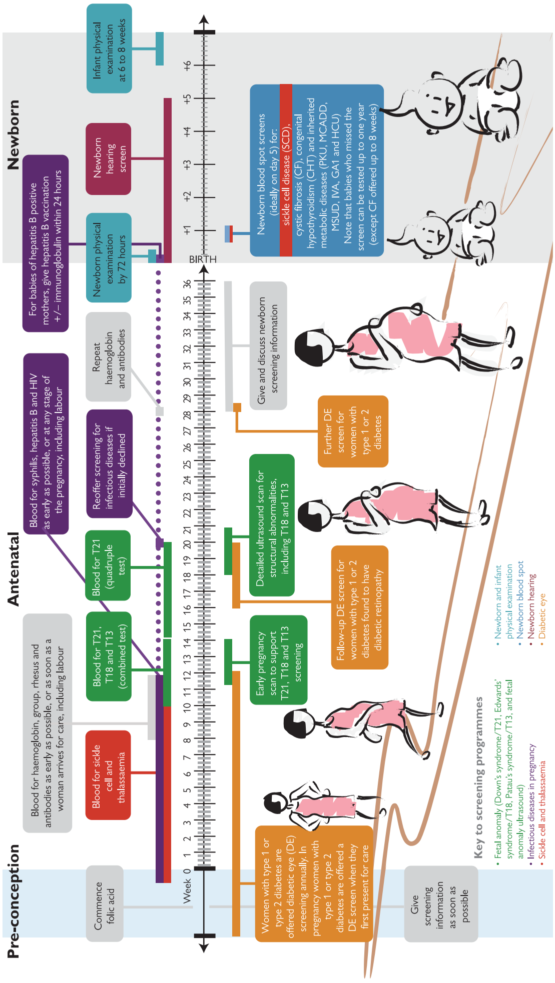
Pre-conception
Antenatal
Newborn
Commence folic acid
Blood for haemoglobin, group, rhesus and antibodies as early as possible, or as soon as a woman arrives for care, including labour
Blood for sickle cell and thalassaemia
Blood for T21, T18 and T13 (combined test)
Blood for T21 (quadruple test)
Blood for syphilis, hepatitis B and HIV as early as possible, or at any stage of the pregnancy, including labour
Reoffer screening for infectious diseases if initially declined
Repeat haemoglobin and antibodies
For babies of hepatitis B positive mothers, give hepatitis B vaccination +/− immunoglobulin within 24 hours
Newborn physical examination by 72 hours
Newborn hearing screen
Infant physical examination at 6 to 8 weeks
Week 0 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30 31 32 33 34 35 36
BIRTH
+1 +2 +3 +4 +5 +6
Women with type 1 or type 2 diabetes are offered diabetic eye (DE) screening annually. In pregnancy women with type 1 or type 2 diabetes are offered a DE screen when they first present for care
Early pregnancy scan to support T21, T18 and T13 screening
Follow-up DE screen for women with type 1 or 2 diabetes found to have diabetic retinopathy
Detailed ultrasound scan for structural abnormalities, including T18 and T13
Further DE screen for women with type 1 or 2 diabetes
Give and discuss newborn screening information
Newborn blood spot screens (ideally on day 5) for: sickle cell disease (SCD), cystic fibrosis (CF), congenital hypothyroidism (CHT) and inherited metabolic diseases (PKU, MCADD, MSUD, IVA, GA1 and HCU)
Note that babies who missed the screen can be tested up to one year (except CF offered up to 8 weeks)
Give screening information as soon as possible
Key to screening programmes
• Fetal anomaly (Down's syndrome/T21, Edwards' syndrome/T18, Patau's syndrome/T13, and fetal anomaly ultrasound)
• Infectious diseases in pregnancy
• Sickle cell and thalassaemia
• Newborn and infant physical examination
• Newborn blood spot
• Newborn hearing
• Diabetic eye

Figure 22.3 Antenatal and newborn screening timeline—optimum times for testing

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Health promotion for pregnant women

Work For most women, work in pregnancy is safe. By law:

- Employers must assess risks to the health/safety of the pregnant woman and adjust for risks accordingly
- Women are entitled to time off work for antenatal care
- Employed women cannot work >33wk into pregnancy unless the woman's GP informs her employer that she may continue
- Employers may not require/allow return to work <2wk after childbirth
- Women who work for an employer qualify for 52wk of maternity leave. Employment rights continue throughout this period. Women can apply for flexible/part-time working hours on return to work

Further information

Health and Safety Executive 🖱 www.hse.gov.uk/mothers

Exercise Moderate exercise is safe and healthy. Advise pelvic floor exercises (➔ p. 821). Avoid:

- Contact sports, high-impact sports, and vigorous racquet sports
- Scuba diving—possible link with fetal birth defects and may cause fetal decompression disease

Drugs Advise women to avoid unnecessary medicines (including OTC) and illicit drugs. Drugs that can be started/continued in pregnancy, if clinically necessary and benefits outweigh risks, include:

- Antacids and ranitidine
- Hormones—levothyroxine, insulin
- Low-dose aspirin (75mg)
- Antihistamines—chlorphenamine
- Metformin
- Laxatives
- Analgesics—paracetamol, codeine-based preparations
- Antihypertensives—methyldopa, nifedipine, labetalol, doxazosin
- Asthma drugs—salbutamol, terbutaline, ipratropium, inhaled steroids
- Antiemetics—cyclizine, domperidone, prochlorperazine, metoclopramide
- Antibiotics—except tetracyclines; avoid trimethoprim in first trimester and at term

Drugs to discontinue or change in pregnancy

- NSAIDs—except low-dose aspirin
- Retinoids, e.g. isotretinoin
- Antibiotics—tetracycline, doxycycline
- Anticonvulsants—take specialist advice
- Antihypertensives—ACE inhibitors, angiotensin receptor blockers
- Warfarin—liaise with obstetrician; may need to change to LMWH

Further information on drugs in pregnancy

BNF 🖱 https://bnf.nice.org.uk/guidance/prescribing-in-pregnancy.html

UK Teratology Information Service ☎ 0344 892 0909 🖱 www.uktis.org

Complementary therapies Use as little as possible.

- Avoid Oil of evening primrose (possible ↑ in PROM)
- No benefit Raspberry leaf tea (but probably no risk either)
- No/limited evidence St John's wort, hypnosis, aromatherapy
- Possibly beneficial Ginger, P6 acupressure and acupuncture for nausea and vomiting; moxibustion for breech presentation; acupuncture for backache/pelvic pain; acupuncture for insomnia

Smoking Stress benefits of quitting at any stage—➔ p. 156. Halving the number of cigarettes smoked results in an average 92g ↑ in birth weight.

Alcohol ➔ p. 762

Diet Normal weight gain in pregnancy is 7–8kg. Do not routinely weigh unless worries about nutrition and/or weight.

Foods to avoid ➔ p.763

Gardening and changing cat litter ➔ p. 763

Travel

- Car Seat belt should go above and below—*not* across—the bump
- Air Check specific requirements of carrier. Most airlines will not accept pregnant women >32wk (rarely, 36wk with a doctor's letter)

> ⚠ Travel involving long periods of immobility (>3h) is associated with ↑ risk of venous thromboembolism. Advise women to drink plenty of non-alcoholic fluids; keep their legs moving while sitting or walk up and down the aisle; and purchase graduated compression hosiery OTC.

Travel abroad Best time to travel is in the second trimester. Travel to high-risk areas is best postponed/cancelled. Avoid travel to places at altitudes >2500m (↑ risk of IUGR/pre-eclampsia).

Malaria Travel to malaria areas is best avoided. If unavoidable, take precautions to avoid mosquito bites (➔ p. 147). Chloroquine and proguanil can be used in usual doses in pregnancy. Give folic acid 5mg od with proguanil. Consider mefloquine for travel to chloroquine-resistant areas. Avoid atovaquone/proguanil and doxycycline.

Contaminated food/water Risk of listeriosis, toxoplasmosis, and hepatitis E. Avoid travel to hepatitis E areas; hepatitis E carries ~20% death rate if infected in the third trimester. Severe diarrhoea may be harmful to the fetus.

Insurance for travel Ensure adequate cover. Most companies insure pregnant women to 28wk, some to 32wk. Reciprocal arrangements do exist with some other countries but do not cover costs of transport or repatriation.

Vaccines Assess risk/benefit ratio on an individual basis.

- Avoid live vaccines BCG, cholera, measles, mumps, rubella, varicella, smallpox, Japanese encephalitis. ❗ Inadvertent administration has not been shown to cause harm
- Inactivated vaccines Give if needed— pertussis, hepatitis A and B, meningococcal (only if significant risk of infection), inactivated polio (normally avoid), rabies, tetanus/diphtheria, yellow fever (avoid unless high risk)

Routine vaccination in pregnancy

- Influenza Pregnancy ↑ risk of complications and death from seasonal influenza. Offer annual flu vaccination to all pregnant women
- Pertussis To protect neonates from severe pertussis infection, pregnant women are currently also offered immunization with combined diphtheria, tetanus, pertussis, and polio vaccine between 16 and 32wk gestation

Who should deliver where?

All those offering maternity care must give women choices about type of care, place of care and birth, and the information to make those choices avoiding personal bias or preference. Who delivers where ultimately depends on the choice that the woman makes (Table 22.2). *Options*:

- Consultant unit
- Midwife or GP/midwife unit integral with/attached to a consultant unit
- 'Isolated unit'—distant from a specialist unit and staffed by midwives or midwives and GPs
- Home (~1% deliveries in the UK)

Legal position of GPs GPs are often fearful of litigation if women opt for delivery outside a specialist unit. Even women with no risk factors can run into problems—rapid intervention to save life is needed in ~5% deliveries. With midwives taking on increasing responsibility for antenatal care in the community, GP-attended home deliveries are very uncommon, and GPs perceive they lack expertise. This compounds their worry.

The legal position is that

- GPs are responsible only for their own acts or omissions
- Midwives are accountable for their own actions and decisions
- The GP only becomes responsible for a woman's care in labour when the midwife attending seeks his/her advice. The GP is then bound by terms and conditions of service to offer advice (either over the telephone or by attending), whether or not the woman had been accepted for maternity care
- If an accident occurs, the GP would be judged against standards of a colleague of similar skills and training, not a specialist obstetrician

Duties of the GP

- Provision of impartial advice about available services locally
- Discussion of the available options in a way to enable the woman to make an informed choice
- To make arrangements for provision of care

Specialist unit vs community-based care In the UK the perinatal and maternal death rates have fallen as the proportion of hospital births has risen. But this is largely due to improvements in housing, nutrition, and antenatal care rather than having babies in hospital per se. Evidence from countries where home birth is the norm suggests that birth outside the hospital setting is safe for healthy women with low-risk pregnancies.

Table 22.2 Reasons why women choose home or hospital births

Home birth	Hospital birth
• To avoid intervention (31%)	• Safety (84%)
• More in control in familiar surroundings (25%)	• Previous hospital birth (6%)
• Previous home birth (11%)	
• More relaxed at home (10%)	
• Fear of hospitals (10%)	
• Continuity of care with midwife (4%)	

⚠ If a woman decides to deliver away from a specialist unit, she should be informed about:

- The facilities and levels of skill and expertise available where she has chosen to deliver, and
- The facilities and specialist services that are available in a specialist unit but *not* where she has chosen to deliver

Record the discussion in her notes.

Advise to deliver in a consultant unit

At booking if

- Pre-existing medical disorders—epilepsy, DM, cardiac, renal, respiratory, hepatitis B, HIV, active genital herpes, IV drug abuse, history of major gynaecological surgery, or known uterine abnormality
- Familial disorder with a high risk of transmission
- ↑ BP
- Height <150cm and primigravida
- Weight at first examination <50kg or >100kg
- Past obstetric history of:
  - Perinatal death
  - Rhesus isoimmunization
  - Pre-eclampsia or eclampsia
  - Antepartum haemorrhage
  - IUGR
  - Caesarean section
  - Postpartum haemorrhage
  - Retained placenta
  - Inverted uterus
  - Shoulder dystocia

If any of the following develop during pregnancy

- Polyhydramnios
- Malpresentation
- Antepartum haemorrhage
- Prolonged pregnancy (>40wk + 10d)
- Preterm labour <37wk
- Suspected IUGR
- Pregnancy-induced ↑ BP
- Multiple pregnancy

Refer to obstetrics to discuss place of delivery if

- Primigravida <18y and >35y or ≥ para 6
- Excessive maternal weight ↑
- Failure of engagement of the head near term in a primigravida
- Past history of prolonged labour, large baby, subfertility, or cone biopsy

⚠ Other rarer medical or obstetric conditions may require specialist advice—if in doubt refer.

Further information

NICE (2008, updated 2019) Antenatal care: routine care for healthy pregnant women. 🖱 www.nice.org.uk/guidance/cg62

Information and support for pregnant women

Emma's diary 🖱 www.emmasdiary.co.uk
National Childbirth Trust (NCT) ☎ 0300 330 0700 🖱 www.nct.org.uk
Ready, Steady, Baby 🖱 www.readysteadybaby.org.uk
Which? Birth Choice 🖱 www.which.co.uk/birth-choice
Your pregnancy and baby guide 🖱 www.nhs.uk/conditions/pregnancy-and-baby

Screening in pregnancy

Most women undergo some form of screening before/during pregnancy aiming to identify, prevent, and treat actual or potential problems (Figure 22.3, ➔ p. 777). Offer women and their partners unbiased information verbally and in writing regarding tests, the meaning and consequences of results, and further options for management. The right to accept or decline should be made clear—and the decision recorded in the antenatal notes.

Pre-pregnancy genetic screening There are many inherited diseases. Most tests give no absolute 'yes' or 'no' but are a risk assessment. Refer couples for genetic screening before pregnancy if they want referral and have factors which put them at high risk of having a baby with a genetic disorder.

Personal or family history of genetic abnormality e.g.:

- Cystic fibrosis
- Down's syndrome
- Sickle cell disease
- β-thalassaemia
- Haemophilia
- Fragile X syndrome
- Polycystic kidneys
- Huntington's chorea
- Duchenne and other muscular dystrophies
- MCAD deficiency

High-risk ethnic groups e.g.:

- Afro-Caribbean origin—sickle cell anaemia
- Indian subcontinent, Far East, southern Europe—thalassaemia
- Ashkenazi Jew—Tay–Sachs disease

❗ The Family Origin Questionnaire (available from 🖱 www.gov.uk) identifies those at risk of carrying a haemoglobinopathy. A blood test establishes carrier status and risk to the baby.

Older women ↑ risk of chromosomal abnormalities including Down's syndrome (➔ p. 841).

Consanguineous couples First-degree cousins who have a baby together have an ↑ risk of congenital malformations in their offspring.

Basic antenatal screening tests Blood and urine tests. Ensure women are given information about the reasons for, significance of, and results of routine tests and record in the notes that permission has been given to do them. Usual tests are:

- Hb estimation (➔ p. 800)
- Blood group
- Antibody screen (➔ p. 770)
- Haemoglobinopathy screen (➔ p. 785)
- Down's syndrome screen (➔ p. 784)
- Urine dipstick for proteinuria (pre-eclampsia screen ➔ p. 786)
- MSU for M,C&S (➔ p. 798)
- Hepatitis B status (➔ p. 797)
- HIV status (➔ p. 785)
- Syphilis status (➔ p. 798)
- Rubella susceptibility

❗ Rubella immune status is not strictly a screening test for this pregnancy but does identify susceptible women (~2.5%) so that postpartum vaccination may protect *future* pregnancies.

Fetal growth and position Measure and record symphysis-fundal height (SFH) at each antenatal appointment from 24wk. Assess fetal presentation by abdominal palpation at ≥36wk. Confirm suspected malpresentation by USS.

Ultrasound scan (USS)

- **Early USS** Offered to all pregnant women at 10wk–13wk 6d for accurate gestational age assessment
- **High-resolution 'anomaly' scan (including fetal echocardiography)** Offered at 18wk–20wk 6d to detect fetal structural abnormalities (Table 22.3). Detection rates vary according to the abnormality, e.g. subtle heart anomalies are less likely to be detected than gross CNS anomalies

Placenta praevia Most low-lying placenta detected at the routine 18–20wk anomaly scan resolve by the time the baby is born. Women with placenta praevia extending over the internal cervical os are offered a further transabdominal USS at 32wk. If this is unclear, transvaginal USS is offered.

Chorionic villus sampling (CVS) Used to detect genetic/metabolic abnormality in high-risk pregnancies. Performed from 11wk–13wk 6d gestation. The developing placenta is sampled per abdomen with USS guidance.

- **Advantages** Undertaken earlier than amniocentesis to allow termination of affected pregnancies at an earlier stage
- **Risks** ~2–4% miscarry; limb defects (💣)

Amniocentesis Sampling of amniotic fluid via transabdominal needle under USS guidance >15wk gestation. 1.9% risk of pregnancy loss. Offered if:

- A high-risk result is obtained following first/second-trimester screening for Down's syndrome or other genetic/chromosomal abnormality
- The woman has had a previous pregnancy affected by fetal anomaly
- There is a strong family history of an inherited disorder

Fetoscopy Fibreoptic visualization of the fetus. Carried out from ~18wk. Enables detection of external abnormalities, fetal blood sampling, and organ biopsy. Fetal loss rate ~4%.

Table 22.3 Fetal anomalies found at the mid-pregnancy scan

Anomaly	Chance of detection at USS (%)
Anencephaly	98
Gastroschisis	98
Edward's syndrome (trisomy 18)	95
Patau's syndrome (trisomy 13)	95
Open spina bifida	90
Bilateral renal agenesis	84
Exomphalos	80
Cleft lip	75
Diaphragmatic hernia	60
Lethal skeletal dysplasia	60
Serious cardiac abnormalities	50

Further information

NICE (2008, updated 2019) Antenatal care: routine care for healthy pregnant women 🖱 www.nice.org.uk/guidance/cg62

RCOG (2010, updated 2017) Amniocentesis and chorionic villus sampling. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg8

Specific antenatal screening tests

Routine antenatal screening is NOT recommended for

Vaginal/genital infections

- Bacterial vaginosis
- Chlamydia (unless <25y ➔ p. 796)
- Group B streptococci
- Genital herpes

Genetic conditions Refer for screening only if a family member is affected:

- Cystic fibrosis
- Fragile X
- Familial dysautonomia

Other infections

- Cytomegalovirus
- Hepatitis C
- Toxoplasmosis

Others

- Thrombophilia
- Thrombocytopenia
- Preterm labour
- Domestic violence

Down's syndrome screening Down's syndrome is the most common single cause of learning difficulty in children of school age. *Incidence*: 1.2/1000 live births. Incidence ↑ with age of the mother (Table 22.4). Screening should detect >75% of Down's syndrome pregnancies, with a screen +ve rate of <3%.

Combined screening test Performed from 10wk–14wk 1d gestation. Screening test of choice, as aids early diagnosis and can be completed in one stage without the need for re-attendance. Produces a single estimate of the woman's risk of having a child with Down's syndrome using:

- Maternal blood tests For human chorionic gonadotrophin (hCG) and pregnancy-associated paraprotein A (PAPP-A), *and*
- Nuchal translucency (NT) USS measurement of the translucency of the nuchal fold in the neck of the fetus done from 11wk 2d–14wk 1d gestation

Quadruple screening test Performed from 14wk 2d–20wk; used for women who book later in pregnancy (~15%). Maternal blood is tested for alpha-fetoprotein (AFP), hCG, unconjugated estriol (uE3), and inhibin A.

Test results Are expressed as a risk assessment (e.g. 1 in 300 at term) or as a +ve or –ve result. A +ve result implies a risk >1 in 150 at term. Ensure all women with +ve results are referred promptly for counselling about further investigation (CVS/amniocentesis) and pregnancy options.

Non-invasive prenatal screening (NIPT) (also known as cfDNA screening) New test that analyses cell-free maternal and fetal DNA in the mother's blood. Highly sensitive (>99%). May be offered in private clinics but currently not available in the NHS except as part of a research/pilot study.

Patau's and Edward's syndrome The combined screening test at 10wk–14wk 1d can be used to screen for trisomy 13 (Patau's syndrome) and trisomy 18 (Edward's syndrome) as well as Down's syndrome. For women booking later in pregnancy, the quadruple screening test only tests for Down's syndrome but most babies with Edward's or Patau's syndrome have abnormalities detected on the routine anomaly scan at 18wk–20wk 6d. NIPT (available privately) may also be used to screen for Patau's and Edward's syndromes.

Anaemia ➔ p. 800 **Rhesus incompatibility** ➔ p. 770

Table 22.4 Levels of risk of having a Down's syndrome pregnancy in relation to a woman's age

Woman's age (y)	Risk as a ratio
20	1:1500
30	1:800
35	1:270
40	1:100
≥45	≥1:50

⚠ The National Screening Committee has recommended that all pregnant women, irrespective of age, should be offered screening for Down's syndrome

Haemoglobinopathy Screening for sickle cell disease and thalassaemia should be offered to all women as early as possible in pregnancy (ideally <10wk). In both cases, women identified as being a carrier (having a trait) or having the disorder should be referred promptly for specialist counselling. Their partners should also be offered screening.

Sickle cell disorders Screening policy depends on prevalence:

- High-prevalence areas (>1.5/10,000 pregnancies) All women are offered screening for sickle cell and other haemoglobin variants
- Low-prevalence areas The Family Origins Questionnaire (available from 🖱 www.gov.uk) is used to assess the risk of either the woman or her partner being a carrier for sickle cell and other haemoglobin variants. Those in identified high-risk groups are offered laboratory testing

Thalassaemia All women are offered FBC early in pregnancy. One of the routine FBC indices, mean cell haemoglobin (MCH) is used as a screening test for thalassaemia. If MCH is ≤27pg, a more diagnostic test using HbA2 (Hb alpha 2) chromatographic analysis is required.

Tay–Sachs disease Genetic condition carried by 1 in 25 Ashkenazi Jews. Offer genetic screening *whether or not* there is a family history. Offer screening for other diseases commonly carried in this population (e.g. Gaucher's disease, familial dysautonomia, cystic fibrosis, Canavan's disease) *only* if there is a family history.

Hepatitis B ➔ p. 797 **Syphilis** ➔ p. 798

Chlamydia screening Is not part of routine antenatal screening, but women under the age of 25y are at high risk of chlamydia infection and may be offered screening through the National Chlamydia Screening Programme (➔ p. 796).

Antenatal HIV testing HIV testing is now offered at the booking appointment as a routine part of antenatal screening in the UK. If the woman screens negative but is at continued high risk of acquiring HIV, a repeat test may be offered later in pregnancy.

❗ If a woman screens +ve for HIV, her partner and any other children in the family should also be offered screening.

Benefits of screening Without intervention, ~25–30% babies born to mothers infected with HIV will become infected with HIV themselves. Avoidance of breastfeeding, antiretroviral therapy, and appropriate management of delivery ↓ risk of transmission to <1%.

If the screening test is declined Document refusal in the woman's notes, explore the reasons for refusal and offer screening again at 28wk.

Screening for other infections Pregnant women who are HIV +ve should be offered screening for genital infection—chlamydia, gonorrhoea, and bacterial vaginosis—done as early as possible in pregnancy and at around 28wk, as co-infection is common in certain subgroups of these women and can ↑ rate of mother-to-child transmission, as well as adversely affecting the pregnancy itself. Also check hepatitis B and C serology unless already done.

Asymptomatic bacteriuria Detection and treatment ↓ the risk of pyelonephritis—➔ p. 798.

Gestational diabetes Identify women at high risk of gestational DM at the booking appointment.

Previous gestational DM or PCOS Screen with early self-monitoring of blood glucose or offer a 2h 75g oral glucose tolerance test (OGTT) at 16–18wk, followed by a repeat OGTT at 28wk if the first test is normal.

Other risk factors include:

- Previous macrosomic baby (≥4.5kg)
- BMI >30kg/m^2
- DM in a first-degree relative
- Family origin associated with high prevalence of DM: South Asian (especially India, Pakistan, or Bangladesh); black Caribbean; Middle Eastern (especially Saudi Arabia, United Arab Emirates, Iraq, Jordan, Syria, Oman, Qatar, Kuwait, Lebanon, or Egypt)

Offer an OGTT at 24–28wk.

Pre-eclampsia Check BP and urinalysis for proteinuria at each antenatal appointment to screen for pre-eclampsia (➔ p. 804).

First contact At first contact, assess the pregnant woman's level of risk for pre-eclampsia. Risk factors for developing pre-eclampsia are:

- Nulliparity or pregnancy interval of >10y
- BMI ≥30kg/m^2 at first contact
- Age ≥40y
- Multiple pregnancy
- Family (e.g. mother, sister) or past history of pre-eclampsia
- Pre-existing vascular (e.g. hypertension, DM) or renal disease

Consider ↑ frequency of BP/proteinuria monitoring in pregnancy for these women—although optimum frequency of BP checks is unclear.

⚠ Warn all pregnant women of the symptoms of advanced pre-eclampsia:

- Headache
- Problems with vision, e.g. blurring or flashing before the eyes
- Bad pain just below the ribs
- Vomiting
- Sudden swelling of face, hands, or feet

If a pregnant woman experiences any of these symptoms in pregnancy she should seek advice from a doctor or midwife as soon as possible.

Psychiatric illness At first contact, ask about:

- History of mental health problems, e.g. schizophrenia, bipolar disorder, severe depression
- Previous treatment from a specialist mental health team (including inpatient care)
- Family history of perinatal mental illness

❗ Ensure that information about any relevant history of mental illness is included in your referral letter for antenatal care.

At booking Ask:

- During the past month, have you often been bothered by feeling down, depressed, or hopeless?
- During the past month, have you often been bothered by having little interest or pleasure in doing things?
- If the woman answers 'yes' to either question, ask: is this something you feel that you need help with?

If a mental health problem is detected, further assessment (e.g. with PHQ-9—➔ p. 979) is indicated.

Postnatal screening ➔ p. 819

Further information

BHIVA (2019 update) Management of HIV in pregnant women 2018 🖱 https://www.bhiva.org/pregnancy-guidelines

NICE (2008, updated 2019) Antenatal care: routine care for healthy pregnant women 🖱 www.nice.org.uk/guidance/cg62

NICE (2014, updated 2018) Antenatal and postnatal mental health: clinical management and service guidance. 🖱 www.nice.org.uk/guidance/cg192

NICE (2015) Diabetes in pregnancy: management from preconception to the postnatal period. 🖱 www.nice.org.uk/guidance/ng3

Public Health England (2013) Fetal anomaly screening: programme overview. 🖱 www.gov.uk/guidance/fetal-anomaly-screening-programme-overview

Public Health England (2013) Sickle cell and thalassaemia screening: programme overview. 🖱 www.gov.uk/guidance/sickle-cell-and-thalassaemia-screening-programme-overview

Public Health England (2016) Infectious diseases in pregnancy screening: programme overview. 🖱 www.gov.uk/guidance/infectious-diseases-in-pregnancy-screening-programme-overview

Public Health England (2019) Family origin questionnaire: sickle cell and thalassaemia screening. 🖱 www.gov.uk/government/publications/family-origin-questionnaire-sickle-cell-and-thalassaemia-screening

Information and support for prospective parents

Antenatal results and choices (ARC) ☎ 0845 077 2290 🖱 www.arc-uk.org

Down's Syndrome Association ☎ 0333 1212 300 🖱 www.downs-syndrome.org.uk

Genetic Alliance Group ☎ 020 7831 0883 🖱 www.geneticalliance.org.uk

Shine (spina bifida and hydrocephalus) 🖱 www.shinecharity.org.uk

Sickle Cell Society ☎ 020 8963 7794 🖱 www.sicklecellsociety.org

SOFT UK (trisomy 13 and 18) 🖱 www.soft.org.uk

UK Thalassaemia Society 🖱 www.ukts.org

Common symptoms in pregnancy

Acute abdominal pain ➔ p. 1078

Backache Affects 60% of pregnant women—usually from the second trimester onwards and worse in the evenings—may interfere with sleep/ activities. Encourage light exercise (unless contraindicated, e.g. pre-eclampsia)—special land and water-based classes are run for pregnant women. Treat with simple analgesia, physiotherapy ± massage.

Breast soreness Most common early in pregnancy. Good support bras are essential (can be purchased from specialist clothing stores). Nipples enlarge and darken at ~12wk.

Carpal tunnel syndrome Affects ~28% pregnant women—➔ p. 460. Reassure usually resolves after pregnancy. Night splints may help. If severe, consider steroid injection. Diuretics do not help. If does not resolve after pregnancy, refer for orthopaedic assessment.

Constipation Affects up to 40% of pregnant women. ↑ fluid and fibre intake. If necessary use a bulk-forming laxative, e.g. ispaghula husk. Avoid bowel stimulants (e.g. senna) as they ↑ uterine activity.

Cramp Leg cramp affects 1 in 3 in late pregnancy. Worse at night. Raising the foot of the bed by 20cm (e.g. 1–2 bricks) can help.

Fatigue

- Early pregnancy Almost universal symptom. Reaches peak at 12–15wk. Advise rest and adjustment of lifestyle. Reassure
- Late pregnancy Due to ↑ physical effort needed to do everyday tasks and sleep deprivation. Check not anaemic, else reassure

Haemorrhoids Affect ~8% of women in the third trimester. May be associated with itching, pain, and bleeding. Advise ↑ fibre intake. Treat prolapse with ice packs and replacement. Topical haemorrhoid applications are commonly used but lack evidence of safety or efficacy.

Headache Usually tension headache. Check BP and urine for protein to exclude pre-eclampsia (➔ p. 804). Treat with rest and analgesia. Migraine may ↑ or ↓ in pregnancy.

Heartburn Affects ~70% of women in the third trimester of pregnancy. Reassure not harmful. Advise low-fat, bland food, small portions, and frequent meals. Avoid eating late at night if worse at night, and consider raising the head of the bed (1–2 bricks under the bed). Avoid gastric irritants, e.g. caffeine. Antacid preparations, e.g. magnesium trisilicate, are helpful if lifestyle modifications are ineffective but may worsen constipation.

> ⚠ Pre-eclampsia can present with epigastric pain—check BP and urine for protein if epigastric/right upper quadrant pain unresponsive to simple antacids. Refer for same-day assessment even if BP is normal and no/ trace proteinuria (➔ p. 804).

Hypotension Common symptom of early pregnancy. Check no bleeding. Advise to avoid standing suddenly and avoid hot baths.

Insomnia Avoid drug treatment. Reassure. Relaxation techniques and mild physical exercise prior to sleep can help.

Itching/pruritus ➲ p. 791

Nausea and vomiting >80% from 4–6wk— ~½ vomit. Occurs at any time of day ('morning' sickness in <20%) and made worse by odours associated with preparation/sight of food. If severe exclude multiple pregnancy, trophoblastic disease, and UTI. Symptoms usually improve by 14–16wk although persist in some.

Management Reassure—normal part of pregnancy. Adjust lifestyle, e.g. ask partner to do the shopping. Advise frequent small meals—avoid greasy/spicy foods, eat foods can face (varies). Maintain fluid intake—small amounts frequently. Self-help measures include ginger and P6 acupressure. If severe/disabling consider antiemetics, e.g. cyclizine 50mg tds. Suppositories are an effective method of administration if PO route is not tolerated. If dehydrated or >2–5kg weight loss (*hyperemesis gravidarum*—1% pregnancies) admit for rehydration.

Peripheral paraesthesia Abnormalities of sensation (e.g. tingling, pins and needles) of hands/feet are common. Reassure. Symptoms usually resolve after delivery. Carpal tunnel syndrome—➲ p. 460.

Skin changes Pigmentation (e.g. linea nigra); spider naevi; abdominal striae; chloasma/melasma; palmar erythema. *Skin rashes*: ➲ p. 791.

Sweating and feeling hot Common. Check apyrexial. If apyrexial, reassure normal in pregnancy. If pyrexial, look for a source of infection.

Swelling Fluid retention affects 80%—ankles, hands/fingers, face. If severe/sudden ↑ in oedema, exclude pre-eclampsia (check BP, dipstick urine for protein).

Symphysis pubis dysfunction 3%. Symphysis separates causing discomfort/pain in lower abdomen/pelvic area radiating to lower back, upper thighs, and perineum. Pain is constant and worse on movement and resolves on rest. Treat with simple analgesia. Consider referral to physiotherapy for pelvic support belt or elbow crutches. Advise rest in a semi-recumbent position when in pain. Generally resolves after delivery but if persists refer to orthopaedics.

Urinary frequency Check MSU—UTI is common in pregnancy and associated with premature delivery—➲ p. 798.

Vaginal bleeding Early pregnancy—➲ p. 766; antepartum haemorrhage—➲ p. 810.

Vaginal discharge Usually ↑ in pregnancy. Investigate if smelly, itchy, sore, or associated with dysuria.

Varicose veins Cause aching legs, fatigue, itch, and ankle/foot swelling. If ankles are swollen, exclude pre-eclampsia (check BP, dipstick urine for proteinuria). Elevate legs when sitting, provide support stockings, and encourage walking/discourage standing still. Complications include *thrombophlebitis*—treat with ice packs, elevation, support stockings, and analgesia—and DVT (➲ p. 802).

Pruritus and rashes in pregnancy

⚠ At booking

- Enquire if the woman has had chickenpox and/or shingles in the past. If not, advise her to make urgent contact if she develops a chickenpox-type rash or has contact with chickenpox or shingles
- Advise the woman to inform the midwife/GP urgently if she develops any rash during pregnancy or has contact with anyone who has a rash

Contact with rashes in pregnancy Many pregnant women have young children and contact with children with rashes is common. *Management*—Figure 22.4.

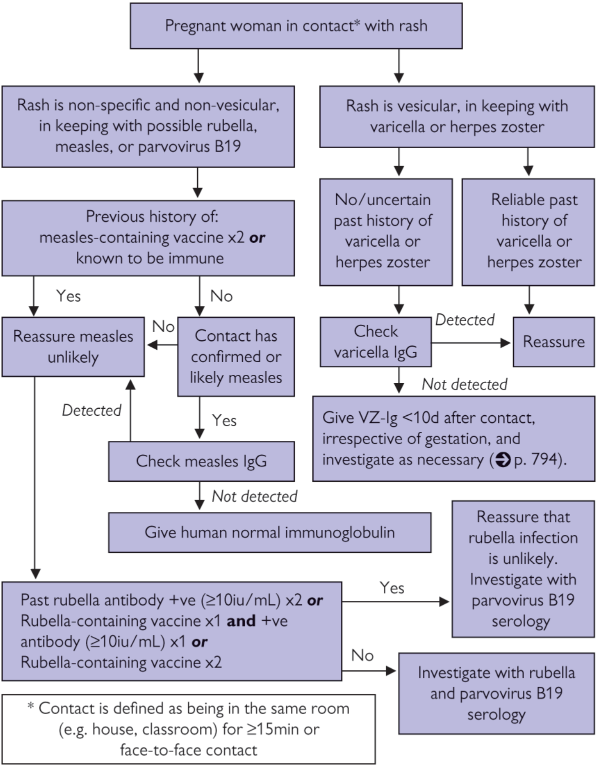

Pregnant woman in contact* with rash
Rash is non-specific and non-vesicular, in keeping with possible rubella, measles, or parvovirus B19
Rash is vesicular, in keeping with varicella or herpes zoster
Previous history of: measles-containing vaccine x2 or known to be immune
Yes
No
No/uncertain past history of varicella or herpes zoster
Reliable past history of varicella or herpes zoster
Reassure measles unlikely
No
Contact has confirmed or likely measles
Check varicella IgG
Detected
Reassure
Not detected
Detected
Yes
Give VZ-Ig <10d after contact, irrespective of gestation, and investigate as necessary (➔ p. 794).
Check measles IgG
Not detected
Give human normal immunoglobulin
Reassure that rubella infection is unlikely. Investigate with parvovirus B19 serology
Yes
Past rubella antibody +ve (≥10iu/mL) x2 or Rubella-containing vaccine x1 and +ve antibody (≥10iu/mL) x1 or Rubella-containing vaccine x2
No
Investigate with rubella and parvovirus B19 serology
* Contact is defined as being in the same room (e.g. house, classroom) for ≥15min or face-to-face contact

Figure 22.4 Investigation of pregnant women in contact with non-specific, non-vesicular rash, vesicular rash, or known cases of measles, rubella, parvovirus varicella, or herpes zoster

Presentation with itching If a woman presents with itching, look for a rash. If there is *no* rash consider:

Hepatic causes Pruritus gravidarum or recurrent cholestasis of pregnancy affects 2–20 in every 100 pregnancies and sometimes runs in families. Usually begins in the third trimester reaching a peak in the last 1mo.

- **Frank jaundice** Rare—refer urgently to an obstetrician
- **No jaundice** Check LFTs. Refer to obstetrics if abnormal. Otherwise, treat with moisturizers (e.g. Diprobase®) ± oily calamine. Antihistamines do not help

Pruritus gravidarum disappears after delivery but recurs in subsequent pregnancies (40–50%) and may recur with CHC.

Other causes

- **Endocrine** DM, thyrotoxicosis, hypothyroidism—consider checking fasting blood glucose and TFTs
- **Renal** Chronic renal failure—check U&E and Cr
- **Haematological** Iron deficiency—check FBC and ferritin
- **Drug allergies**
- **Psychological** Obsessive states, schizophrenia

❗ If no cause is found and the problem persists, refer to obstetrics.

Presentation with a rash Consider:

- Common skin diseases, e.g. eczema, psoriasis, urticaria
- Skin changes specific to pregnancy
- Infectious causes:
  - Rubella
  - Parvovirus B19
  - Chickenpox/shingles
  - Measles
  - Streptococcal infection
  - Meningococcal infection
  - Enterovirus infection
  - EBV
  - CMV
  - Syphilis

Itchy rashes specific to pregnancy

- **Abdominal striae** May itch
- **Pruritic urticarial papules and plaques of pregnancy (PUPPP)** 1 in 240 pregnancies. Occurs in first/multiple pregnancies or if excessive weight ↑ in pregnancy. Intensely itchy rash usually confined to lower abdomen/buttocks. Appears at >35wk gestation. Treat with calamine and/or topical steroids. Clears spontaneously <6wk (often days) after delivery. Recurrence in subsequent pregnancies is rare
- **Pemphigoid gestationis** Rare. Starts in mid-pregnancy and appears as a generalized, intensely itchy rash. Refer for specialist management. May recur in subsequent pregnancies/with CHC
- **Impetigo herpetiformis** Rare. Starts in the third trimester. Mild itch. Systemically unwell. Refer. Remits after delivery but may recur in later pregnancies

Pregnant women with rash illness See Figure 22.5, ➔ p. 793.

Further information

Public Health England (2019) Rash in pregnancy. 🖱 www.gov.uk/government/publications/viral-rash-in-pregnancy

RCOG (2011, updated 2014) Obstetric cholestasis. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg43

Rash illness in pregnancy

Rubella Presents with fever, LNs (including suboccipital nodes), and a pink maculopapular rash which lasts 3d. 50% of mothers infected with rubella are asymptomatic. Incubation is 14–21d and once infected, patients are infectious from 7d before the rash appears until 7d after. Asymptomatic reinfection of women who have received vaccination can also occur, so serology is essential in all pregnant rubella contacts.

Risk to the baby Abnormalities that can occur as a result of *in utero* infection include: cataract, deafness, cerebral palsy, mental retardation, microcephaly, microphthalmia. If the mother is infected with rubella at >20wk gestation, infection does not affect the baby.

Transmission risk to the baby

- <11wk gestation 90% have adverse outcome
- 11–16wk gestation 20% have adverse outcome
- >16wk gestation Minimal risk of deafness only

Transmission risk is much lower with reinfection (<5%). There is no treatment to prevent transmission.

Management

- Contact with a non-vesicular rash or rubella— see Figure 22.4, ➲ p. 790
- Presentation with a non-specific, non-vesicular rash or suspected rubella infection—send blood for serology for rubella and parvovirus B19 (Figure 22.5). Refer if proven infection. After further investigation and discussion of risks, women infected with rubella at <20wk may be offered termination of pregnancy

Vaccination after pregnancy Give postnatal MMR vaccination routinely to all women found not to be rubella immune in pregnancy.

Parvovirus B19 infection Febrile illness often accompanied by tenderness of the joints/arthritis affecting hands, wrists, and knees—usually lasts 1–2wk. There may be a fine rash over the trunk and extremities. Infectious from 10d before rash appears. Incubation period 13–18d.

Parvovirus B19 in pregnancy ~50% of young women in the UK are not immune. Risk of infection in pregnancy ~1 in 400. Risk of a non-immune mother contracting the infection from a child who has Fifth disease (slapped cheek) is ~50%.

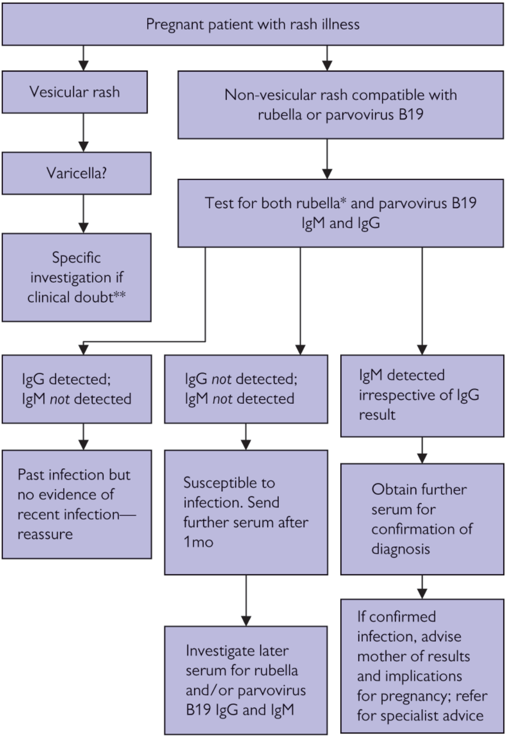
Risk to the baby If infection is at <20wk gestation, there is a 9% ↑ miscarriage rate. 3% (14–56 babies/y in the UK) develop hydrops fetalis >3wk after infection due to anaemia—½ of those babies die. There are no long-term effects from an infection which does not cause miscarriage or hydrops.

Transmission rate to the baby Depends on gestation at the time of infection. There is no treatment to prevent transmission.

- <4wk gestation There is no transmission
- From 5–16wk gestation Transmission rate ~15%
- >16wk gestation 25–70% transmission rate (↑ with gestation)

Management

- Contact with a non-vesicular rash or parvovirus—see Figure 22.4, ➔ p. 790
- Presentation with a non-specific, non-vesicular rash or suspected parvovirus infection—send blood for serology for rubella and parvovirus B19 (Figure 22.5). Refer if proven infection. USS surveillance is started 4wk after onset of illness or seroconversion and then every 1–2wk until 30wk. If there are any signs of hydrops fetalis on USS, the mother is referred to a regional centre for consideration of intrauterine transfusion. Early transfusion ↑ chances of the baby's survival


Pregnant patient with rash illness
Vesicular rash
Non-vesicular rash compatible with rubella or parvovirus B19
Varicella?
Test for both rubella* and parvovirus B19 IgM and IgG
Specific investigation if clinical doubt**
IgG detected; IgM *not* detected
IgG *not* detected; IgM *not* detected
IgM detected irrespective of IgG result
Past infection but no evidence of recent infection—reassure
Susceptible to infection. Send further serum after 1mo
Obtain further serum for confirmation of diagnosis
Investigate later serum for rubella and/or parvovirus B19 IgG and IgM
If confirmed infection, advise mother of results and implications for pregnancy; refer for specialist advice

* Irrespective of past testing or immunization.

** Confirm by detection of varicella virus, antigen, or DNA in vesicle fluid.

Figure 22.5 Investigation of pregnant women with a rash illness

Source: data from http://hpa.org.uk

Cytomegalovirus (CMV) More frequent cause of birth defect than rubella in the UK—5 in 1000 live births—10% develop handicap. The fetus is most vulnerable when infection occurs in early pregnancy. Maternal disease may be asymptomatic or a mild flu-like illness. Occasionally there is a rash. No effective prevention strategy.

Measles Much rarer in the UK since introduction of routine MMR vaccination for children. Presents with coryza, lymphadenopathy, conjunctivitis, and disseminated maculopapular rash which becomes confluent. Complications include pneumonia, otitis media, and encephalitis. Infection in pregnancy can lead to intrauterine death and preterm delivery. There are no associations with congenital infection or abnormalities.

Exposure to possible measles infection See Figure 22.4, ➲ p. 790.

Treatment of neonates Human normal immunoglobulin (HNIG) is given to neonates born to mothers with a measles rash that appears from 6d before to 6d after birth.

Vaccination after pregnancy Give postnatal MMR vaccination routinely to all women found not to be measles-immune in pregnancy.

Chickenpox Contact with chickenpox in pregnancy is common. People with chickenpox are infectious from 2d before the rash appears until the rash has finished cropping and crusted over. Incubation period is 14–21d.

Pre-conceptual prevention An effective chickenpox vaccine is available in the UK. There is no UK policy to screen women for immunity to varicella infection, however it is possible to check immune status and vaccinate non-immune women prior to pregnancy. Pregnancy should be avoided for 3mo after immunization.

Exposure in pregnancy If the mother has definitely had chickenpox there is no risk to herself or the baby. If she does not recall having chickenpox, check her immunity—80% have antibodies from silent infection (Figure 22.4, ➲ p. 790).

In cases of 'at risk' exposure Arrange for VZ-Ig to be given to mother and/or baby. This can be lifesaving and significantly ↓ disease severity if given ≤10d after exposure. Babies are at risk if:

- The mother develops chickenpox from 7d before to 7d after delivery
- The mother is not immune and the baby is exposed to chickenpox <7d after birth
- The baby has been exposed to chickenpox and has potentially inadequate transfer of maternal antibodies (e.g. preterm babies <28wk, babies weighing <1000g at birth, babies who have had blood transfusions). VZ-Ig can be given without antibody testing to these babies, but where possible, test
- Duration of protection from VZ-Ig is limited. Give a second dose if still at risk and further exposure occurs and ≥3wk since first dose. Check antibody status again before giving second dose

💣 Some advocate use of prophylactic aciclovir for women with significant additional risk factors, e.g. immunosuppression, smokers, women who did not receive early VZ-Ig, or those in the second half of pregnancy.

Risk to the mother Chickenpox infection complicates 2–3 in 1000 pregnancies. Chickenpox pneumonia is more common (10%) and can be severe (1 in 1000 mortality).

Risk to the baby Rates of transmission are higher later in pregnancy (~50% >36wk; 5–10% <28wk). Infection:

- <20wk Causes miscarriage. Fetal varicella syndrome affects 1–2%—segmental skin defects/scarring, limb hypoplasia ± paresis, low birth weight, microcephaly, neurological abnormalities (e.g. hypotonia, eye defects). May occur up to 28wk gestation
- 20–37wk Intrauterine infection or death, shingles in childhood
- 1wk before–1wk after delivery All babies should be given VZ-Ig. Onset 4d before delivery–2d after delivery carries a 20% risk of overwhelming neonatal infection—these babies should be given aciclovir in addition to VZ-Ig. Seek specialist advice

Management If clinical doubt, confirm infection by detection of varicella virus, antigen, or DNA in vesicle fluid (Figure 22.5, ➔ p. 793). Treat with aciclovir 800mg 5×/d PO for 1wk if presents <24h after the rash appears and the mother is >20wk gestation. Monitor daily. If <28wk gestation, refer for detailed USS 5wk after infection to exclude fetal varicella syndrome.

Admit if

- Chest symptoms
- Neurological symptoms other than headache
- Severe disease—dense rash/numerous mucosal lesions
- Significant immunosuppression, e.g. HIV +ve
- Haemorrhagic rash or bleeding

Consider admission if

- Pregnancy approaching term
- Bad obstetric history
- Poor social circumstances
- Unable to monitor the woman closely, e.g homeless, traveller
- Smoker
- Chronic lung disease

Refer for urgent hospital assessment if No deterioration but:

- Fever persists, *or*
- Cropping of the rash continues >6d

❗ Warn pregnant women with chickenpox to avoid contact with anyone potentially at risk of developing severe chickenpox, especially other pregnant women or neonates.

Rash infections that cause no harm to the fetus

- Epstein–Barr virus (EBV)
- Enteroviruses—Coxsackie virus A/B; echovirus; enterovirus 68–71—cause disease, such as hand, foot, and mouth. Some enteroviruses can cause severe neonatal infection, and prophylactic immunoglobulin may be necessary—seek specialist advice

Further information

Public Health England (2019) Rash in pregnancy. 🖱 www.gov.uk/government/publications/viral-rash-in-pregnancy
RCOG (2015, updated 2018) Chickenpox in pregnancy. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg13

Other infections in pregnancy

Bacterial vaginosis Present in ~10% of pregnant women—asymptomatic in half. Associated with ↑ preterm birth (×2). There is no screening policy in the UK, but, if detected:

- Treat with metronidazole PO—400mg bd for 7d or 2g stat
- An alternative is clindamycin 2% cream 5g nocte PV for 1wk

❗ Treatment may not lower the risk to the pregnancy.

Chickenpox ➔ p. 794

Chlamydia ~1 in 20 pregnant women has chlamydia infection. During pregnancy, chlamydia infection is associated with IUGR, preterm birth, and low birthweight. It can also pass to the baby during delivery, causing eye and/or chest infections. Postpartum, chlamydia can cause uterine infection. Treat if detected (➔ p. 716)—follow-up with swabs to confirm eradication. Refer any affected neonates for expert advice.

Screening As part of the National Chlamydia Screening Programme, women <25y can ask for chlamydia self-test kits at a variety of healthcare and community settings. Otherwise there is no evidence of cost-effectiveness of routine antenatal screening for STIs apart from HIV and hepatitis B.

Coughs, colds, and 'flu Little threat to the pregnancy. *Advise*: fluids, paracetamol, and rest. Inhaled decongestants are safe, but avoid cough linctus and OTC composite preparations. Treat any secondary infections as needed.

Cytomegalovirus (CMV) ➔ p. 794 **Enteroviruses** ➔ p. 795

Genital herpes^{G} Affects ~10% of the UK population (diagnosis made in 1 in 3). Risk is greatest if the primary attack occurs at >34wk gestation. Secondary attacks are much less of a problem. Risks include:

- Passing the infection to the baby at the time of delivery
- IUGR (primary infection only)
- Early labour

❗ Elective Caesarean section is advised at term if a primary attack occurs during pregnancy at >28wk gestation. 💣 It is controversial if Caesarean section is preferable if there is an active secondary attack at the time of labour.

Gonorrhoea <1 in 1000 pregnancies. Can pass to the baby during delivery, causing eye infections. Treat if detected—➔ p. 717. Refer affected infants urgently for expert advice. Follow-up with swabs to confirm eradication.

Group B Streptococcus (GBS)^{G} Bacterium carried in the vagina by >1 in 4 pregnant women (20% of non-pregnant women). Usually harmless but if transmitted to the baby during delivery can cause neonatal septicaemia, pneumonia, or meningitis.

Prevention of neonatal infection There is currently no UK screening programme to detect women carrying GBS infection during pregnancy. Treatment with IV antibiotics during labour is advised in 'high-risk' scenarios only:

- Early labour <37wk

- Prolonged (>18h) or early (<37wk) rupture of the membranes
- GBS detected in urine in pregnancy
- If the woman has a temperature >37.8°C during labour
- If a previous baby has been affected with the condition (10× ↑ risk)

❗ If found incidentally during pregnancy (e.g. detected on a swab done for another reason), there is no evidence that treatment is effective. However antibiotics are given during labour.

Hepatitis B Prevalence of HBsAg in pregnancy is up to 1% (depending on geographical area). Women are routinely offered screening for hepatitis B infection in pregnancy. Transmission to the baby occurs during labour (up to 30% of infants of women seropositive for HBsAg and 90% of infants of women seropositive for both HBsAg and HBeAg). Infants infected are at high risk (~90%) of becoming chronic carriers and of developing chronic liver disease ± premature death.

Postnatally Refer infected women for hepatology assessment. Infected mothers should not donate their milk.

Immunization[G] Give hepatitis B vaccine as soon as possible (<24h) after birth to babies born to carrier mothers with the addition of immunoglobulin (HBIG) if the mother carries the hepatitis B e-antigen or had acute HBV infection during pregnancy. 85–95% effective in preventing neonatal hepatitis B infection. Further doses of vaccine are required at 1mo of age. The infant should then have 3 doses of hepatitis B vaccination as part of the routine childhood immunization schedule at 2mo, 3mo, and 4mo of age, and a booster dose at 1y at the same time as follow-up testing.

Hepatitis C Prevalence in pregnant women is 0.14–0.8%. Except when initial infection of the mother occurs during pregnancy (when transfer rate is much higher), transmission rate to the fetus is ~5%. To date there is no evidence that HCV can be transferred to the child by breastfeeding. Infants at risk can be screened for HCV infection at 12mo (RNA screen) or 18–24mo (HCV antibody test). The majority of infants that acquire HCV infection via their mothers develop chronic hepatitis. Treatment is with interferon and achieves viral clearance rates of 40%.

HIV[G] Prevalence of HIV among pregnant women in the UK varies from 0.04% to 0.4% depending on geographical area. Up to 50% of infants of HIV seropositive mothers are pre- or perinatally infected with HIV, accounting for 90% of HIV infections in childhood. Avoidance of breastfeeding, antiretroviral therapy, and appropriate management of delivery ↓ risk of transmission to <1%. A detailed fetal anomaly scan is important if there is first-trimester exposure to antiviral treatment (including folate antagonists) as possible ↑ risk of congenital abnormality. ❗ There is a theoretical concern of mother-to-child transmission with invasive prenatal diagnosis. Take specialist advice.

Antenatal HIV testing ➔ p. 785

Fetal abnormalities include Wide-set eyes, short nose, patulous lips, 'box' forehead, and growth failure. However diagnosis is usually made from 6mo–2y of age when the child presents with lymphadenopathy, recurrent or opportunistic infections, failure to thrive, or progressive encephalopathy. Expert advice is needed throughout pregnancy and for neonatal follow-up.

Box 22.2 Advice for prevention of toxoplasmosis and listeriosis

- Only eat well-cooked meat
- Wash hands, cooking utensils, and food surfaces after preparing raw meat
- Keep raw meat and cooked foods on separate plates
- Wash all soil from fruit and vegetables before eating
- If possible avoid cleaning cat litter (or use gloves and wash hands after)
- Use gloves when gardening and wash hands afterwards

Listeriosis Rare. May occur in epidemics. Infection of the mother is usually via infected food, e.g. pâté, soft cheese, milk (Box 22.2). Detection is with blood cultures. Suspect if unexplained fever >48h, and refer for expert advice.

Maternal symptoms Fever, shivering, myalgia, headache, sore throat, cough, vomiting, diarrhoea, vaginitis.

Consequences Miscarriage (may be recurrent), stillbirth, premature labour, transmission to the fetus (in 2nd/3rd trimester). Infection in the newborn infant manifests in pneumonia ± meningitis.

Malaria Serious complications are more common in pregnancy (cerebral malaria has 50% mortality). Suspect in any pregnant woman who has a fever and has recently visited an infected area. Seek immediate expert advice.

Measles ➔ p. 794 **Parvovirus B19** ➔ p. 792 **Rubella** ➔ p. 792

Syphilis Prevalence 0.07%. ~70–100% of pregnant mothers with primary, untreated syphilis transmit the disease to the fetus (1 in 3 die *in utero*). In the early latent phase, risk of transmission is ~40% and ~10–15% in the late latent phase. Neurological abnormalities as a result of congenital syphilis include encephalopathy and sensorineural deafness. Treatment ↓ risk of transmission by >98%. Refer for specialist assessment if +ve result on routine antenatal VDRL testing. ❗ +ve result is NOT specific to syphilis.

Thrush More common in pregnancy. Not harmful to the fetus. Requires treatment only if causes troublesome itching, soreness, or discharge. *Treatment*: imidazole pessaries for 1wk.

Toxoplasmosis Caused by a parasite found in raw meat and cat faeces. Up to 90% of women have not had toxoplasmosis before pregnancy and ~2 in every 1000 will catch it during pregnancy. 30–40% pass it to their fetus. Infection may result in miscarriage, stillbirth, growth problems, blindness, hydrocephalus, brain damage, epilepsy, or deafness. Risk of transmission to the fetus is related to gestation at the time of infection—third trimester ~70%; first trimester ~15%. If infection is suspected refer for specialist advice. Prevention—Box 22.2.

Urinary tract infections 1 in 25 women develops UTI in pregnancy. If suspected, send MSU to confirm diagnosis and start antibiotics (e.g. cefalexin 250mg tds) immediately. Recurrent UTIs in pregnancy should be investigated—consider renal ultrasound >12wk after delivery.

Screening for UTI Routine screening with MSU for UTI is offered at booking. 2–5% of pregnant women have asymptomatic bacteriuria, defined as pure growth of $>10^5$ organisms/mL—1 in 3 will develop symptomatic infection (acute cystitis, pyelonephritis) if left untreated. Both untreated bacteriuria and frank UTI are associated with preterm delivery and IUGR. Treat for at least 1wk with suitable antibiotic (avoid trimethoprim in first trimester). Check MSU following treatment to ensure infection has cleared.

Whooping cough Newborn babies are at particular risk of severe whooping cough infection. Pregnant women are advised to have whooping cough vaccination from 28–38wk (ideally 28–32wk) gestation to provide passive immunity for their babies.

Zika virus (ZIKV)[G] Spread by mosquito vector or sexually transmitted. Cases in the UK are all imported and develop 3–12d following travel to Zika areas. For most healthy individuals, infection is either asymptomatic or causes a mild, self-limiting flu-like illness (including any of: rash, itching/pruritus, fever, headache, arthralgia/arthritis, myalgia, conjunctivitis, lower back pain, ± retro-orbital pain). Rarely, ZIKV is followed by Guillain–Barré syndrome (➜ p. 516).

❗ Zika virus infection in pregnancy is a cause of congenital brain abnormalities

Prevention of ZIKV infection

- Take precautions to ↓ risk of mosquito bites (➜ p. 147)
- Advise to avoid becoming pregnant while travelling in an area with ↑ Zika risk, and on return for a further:
  - 8wk if only the woman travelled, or
  - 6mo if travelled with partner or just partner travelled
- Advise women already pregnant to use a barrier method of contraception while abroad and for the duration of pregnancy to avoid potential transmission from her partner

Management in pregnancy

- **If no clinical illness** Consistent with ZIKV during travel or <2wk after return, arrange fetal USS as soon as possible, and (depending when in pregnancy baseline scan is), again at 18–20wk and 28–30wk; refer to a fetal medicine specialist if any abnormal USS findings
- **If clinical illness** During travel or <2wk after return consistent with ZIKV, send blood for serology. If +ve, refer for specialist care. If –ve, consider repeating test and, if still –ve, return to usual care

Further information

BHIVA (2019 update) Management of HIV in pregnant women 2018. 🖱 https://www.bhiva.org/pregnancy-guidelines
Public Health England (2017) Green book: hepatitis B. 🖱 www.gov.uk/government/publications/hepatitis-b-the-green-book-chapter-18
Public Health England (2017) Green book: pertussis. 🖱 www.gov.uk/government/ publications/pertussis-the-green-book-chapter-24
RCOG (2014) Management of genital herpes in pregnancy. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/genital-herpes
RCOG (2016, updated 2019) Zika virus infection and pregnancy. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/zika-virus-infection-and-pregnancy
RCOG (2017) Prevention of early onset neonatal group B streptococcal disease. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg36

Information for patients

Group B Streptococcus Support ☎ 01444 416 176 🖱 www.gbss.org.uk

A–Z of medical conditions in pregnancy

Pre-existing disease may affect pregnancy or, conversely, pregnancy may have an impact on pre-existing disease. Common medical conditions are listed in this section. If a woman has a less common condition that might impact or be affected by pregnancy, seek early specialist advice.

Anaemia Defined as Hb <11g/dL early in pregnancy, or <10.5g/dL after 28wk. Common in pregnancy (20%). Some ↓ in Hb is physiological due to ↑ in plasma volume, however iron requirements ↑ ×2–3 and folate ↑ ×10–20 during pregnancy. Anaemia is usually due to iron deficiency. Complications include excessive fatigue and poorer fetal outcome. *Risk factors*:

- Multiple pregnancy
- Frequent pregnancies
- Poor diet
- Starting pregnancy anaemic
- Haemoglobinopathy

Screening Hb is routinely screened at booking and again at 28wk.

Management Routine use of oral iron for all pregnant women is of no proven benefit and may cause harm. Women in high-risk groups (e.g. multiple pregnancy) may routinely be given prophylaxis—follow local policies. If Hb is <11g/dL at booking or <10.5g/dL at 28wk, start iron (e.g. ferrous sulfate 200mg tds) and folic acid (5mg od). Repeat Hb in 2wk. If there is no response to oral iron, exclude occult infection (e.g. UTI); check haematinics; consider referral for parenteral iron.

Antiphospholipid syndrome May be 1° (occurs alone) or 2° to other connective tissue disease (usually SLE). Antiphospholipid antibodies (lupus anticoagulant and/or anticardiolipin antibodies), are present plus a history of:

- Arterial or venous thrombosis, and/or
- Recurrent pregnancy loss (typically second trimester—➲ p. 767)

Specialist referral is essential. Treated with low-dose aspirin + LMWH from 6–34wk.

Asthma Affects ~5% of pregnant women. Generally improves with pregnancy, especially into the third trimester. In most cases, treat asthma as usual—most drugs commonly used for treatment of asthma are safe in pregnancy.

❗ Leukotriene receptor antagonists have limited safety data—stop/change or seek specialist advice. Women with poorly controlled asthma are at risk of early labour and IUGR. Syntometrine for third stage of labour should be avoided as it contains ergometrine which can cause a severe asthma attack. There is a tendency to worsening of asthma after delivery.

Cardiac disease Risk of death is highest where pulmonary blood flow cannot be ↑, e.g. Eisenmenger's syndrome, primary pulmonary hypertension.

Management Specialist obstetric care is needed for all patients with a pre-existing cardiac condition. Where possible refer to a cardiologist before conception for discussion of risks. Antibiotic prophylaxis for delivery may rarely be needed if structural cardiac disease—seek specialist advice.

Murmurs in pregnancy Check heart sounds. Murmurs are common. Consider any heart murmurs detected during pregnancy significant and refer for further evaluation with echocardiography—90% will be physiological.

Depression A significant cause of maternal death.

Pre-existing depression Consider referral for pre-conceptual specialist mental health advice. When antidepressants are being used, weigh up the pros and cons of discontinuing treatment during pregnancy.

Screening for depression Antenatal ➔ p. 787; postnatal ➔ p. 819.

Depression in pregnancy For women with mild/moderate depression, consider self-help strategies and talking therapies (e.g. CBT) first. Monitor regularly using depression questionnaires (e.g. PHQ-9—➔ p. 979). Weigh up risks of antidepressant medication against benefits. Involve specialist community midwife teams and mental health services early.

Postnatal depression ➔ p. 819

Diabetes ➔ p. 806 **Eclampsia** ➔ p. 1084 **Epilepsy** ➔ p. 807
HIV ➔ p. 797 **Hypertension** ➔ p. 804 **Infection** ➔ p. 796

Jaundice Any cause of jaundice may occur in pregnancy. Investigate and treat according to cause. Common causes are: viral hepatitis; gallstones; Gilbert's or Dubin–Johnson syndrome.

Jaundice peculiar to pregnancy

- Cholestasis of pregnancy/pruritus gravidarum—➔ p. 791
- Pre-eclampsia—➔ p. 804
- Severe hyperemesis Jaundice is a complication—➔ p. 789
- Acute fatty degeneration of the liver Rare. Usually >30wk gestation. The mother develops abdominal pain, jaundice, headache, and vomiting. Admit

Pre-eclampsia ➔ p. 804

Renal disease Refer for specialist obstetric care. Pre-eclampsia is more common—monitor carefully and refer early if pre-eclampsia is suspected.

Renal failure If mild renal failure and no ↑ BP, >96% have successful pregnancies without adverse effect. Outcomes are less good the more severe the renal failure with complications including worsening hypertension, pregnancy-related ↓ in renal function, IUGR, and preterm delivery.

Women on dialysis Conception is uncommon. High rate of miscarriage and intrauterine death. ~40–50% live birth rate. Mothers are prone to volume overload, polyhydramnios, and severe ↑ BP ± pre-eclampsia. A 50% ↑ in duration/frequency of dialysis is needed during pregnancy.

Renal transplant Risk of first-trimester miscarriage is ↑ but pregnancies that survive are >90% successful. Immunosuppressant drugs must be continued—they are not harmful to the fetus. Pregnancy does not affect long-term survival of the transplanted kidney. Pelvic position of the transplant does not compromise vaginal delivery.

Rheumatoid arthritis (RA) Symptoms often improve during pregnancy and worsen in the puerperium. Do not use NSAIDs for joint pain >24wk gestation as can result in closure of the fetal ductus arteriosus. Paracetamol or paracetamol + codeine combinations are safe. If a woman taking DMARDs is planning pregnancy, refer pre-conception for specialist medication review.

Disease-modifying drugs

- Sulfasalazine—folic acid supplementation is recommended
- Azathioprine—associated with IUGR
- Penicillamine—may weaken fetal collagen
- Methotrexate and most biological agents are contraindicated

Systemic lupus erythematosus (SLE) Exacerbations are common in pregnancy.

- **Effects on fetus** IUGR; neonatal lupus (from passively acquired maternal antibodies—usually self-limiting skin rash)
- **Effects on mother** Renal complications may worsen and be associated with ↑ BP ± pre-eclampsia; oligohydramnios; premature delivery

Management If planning pregnancy refer for review of drugs. Once pregnant refer for specialist obstetric care. Pain control—as for RA.

Immunosuppressive drugs

- Azathioprine—may cause IUGR
- Hydroxychloroquine—risk of deposits in fetal eye/ear
- Cyclophosphamide and methotrexate are contraindicated

Thyroid disease Refer for specialist obstetric advice.

Hyperthyroidism Usually Graves' disease. Severity ↓ through pregnancy. May be associated with neonatal goitre, hyper- or hypothyroidism. Continue treatment with carbimazole aiming to keep plasma T_4 at the top of the normal range. Propylthiouracil is preferred postpartum if breastfeeding, as less concentrated in breast milk.

Hypothyroidism If untreated associated with infertility, ↑ rate of miscarriage, stillbirth, and fetal abnormality. Levothyroxine dose usually needs to ↑ in pregnancy—the fetus is not affected by maternal thyroxine. Check TFTs in each trimester.

Thromboembolism (VTE)[G] Most common direct cause of maternal death in the UK. Pregnancy ↑ risk of VTE ×10—even in very early pregnancy. *Incidence*: ~1 in 100 pregnancies (20–50% antenatal).

⚠ Suspect DVT and/or PE in pregnancy/puerperium if:

- Leg pain and/or swelling
- Chest pain and/or breathlessness
- Mild unexplained fever

Management of suspected VTE Refer as an emergency for confirmation of diagnosis and initiation of treatment. D-dimer tests are unreliable in pregnancy and should not be used. If there will be delay until specialist assessment, start LMWH in the interim.

Management of confirmed VTE The woman is anticoagulated during the remainder of pregnancy and for ≥6wk postpartum (minimum 3mo total). Avoid warfarin and direct oral anticoagulants (DOACs) during pregnancy—use LMWH instead. Avoid DOACs if breastfeeding but warfarin is safe postpartum and during breastfeeding. Advise women to wear graduated compression hosiery for ≥2y after DVT.

Antenatal prevention of VTE Assess risk of thrombosis:

- High risk Women require thromboprophylaxis with LMWH if:
  - Recurrent venous thromboembolism (VTE), *or*
  - Single previous VTE that is unprovoked or oestrogen related or associated with a personal or family history of thrombophilia
- Intermediate risk May require thromboprophylaxis with LMWH. Seek specialist advice if:
  - Single previous VTE and no personal/family history of thrombophilia
  - Thrombophilia with no history of VTE, *or*
  - Pregnancy with co-morbidities that ↑ VTE risk (e.g. heart/lung disease, surgery, cancer, IV drug misuse, inflammatory conditions, SLE, sickle cell disease, nephrotic syndrome)
- Lower risk If ≥3 risk factors, consider thromboprophylaxis:
  - Age >35y
  - Obesity (BMI >30kg/m^2)
  - Parity ≥3
  - Smoker
  - Gross varicose veins
  - Current systemic infection
  - Pre-eclampsia
  - Immobility (e.g. paraplegia, symphysis pubis dysfunction)
  - Long-distance travel
  - Dehydration, hyperemesis, or ovarian hyperstimulation syndrome
  - Multiple pregnancy or assisted reproduction

Postnatal prevention of VTE Assess risk of thrombosis:

- High risk Women require thromboprophylaxis with LMWH for 6wk postnatally if any previous VTE and required antenatal LMWH
- Intermediate risk Give thromboprophylaxis with LMWH for 7d if:
  - Caesarean section in labour
  - BMI >40 kg/m^2
  - Asymptomatic thrombophilia (inherited or acquired)
  - Prolonged hospital admission, or
  - Medical co-morbidities, e.g. heart/lung disease, SLE, cancer, inflammatory conditions, nephrotic syndrome, sickle cell disease, IV drug use

❗ Extend the period of LMWH use if ongoing risk factors.

- Lower risk If ≥2 risk factors, consider thromboprophylaxis for 7d (longer if >3 risk factors):
  - Age >35y
  - Obesity (BMI >30kg/m^2)
  - Parity ≥3
  - Smoker
  - Gross varicose veins
  - Current systemic infection
  - Pre-eclampsia
  - Long-distance travel
  - Immobility (e.g. paraplegia, symphysis pubis dysfunction)
  - Prolonged labour (>24h)
  - PPH >1L or blood transfusion
  - Mid-cavity rotational operative delivery
  - Elective Caesarean section
  - Any surgical procedure in the puerperium

Further information

NICE (2014, updated 2018) Antenatal and postnatal mental health: clinical management and service guidance. 🖱 www.nice.org.uk/guidance/cg192

RCOG (2015, updated 2018) Thrombosis and embolism during pregnancy and the puerperium: reducing the risk. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg37a

RCOG (2015, updated 2018) Thrombosis and embolism during pregnancy and the puerperium: acute management. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg37b

Hypertension in pregnancy

Chronic hypertension Hypertension present <20wk into pregnancy (or woman taking antihypertensive medication prior to pregnancy). More common in older mothers. May worsen in later pregnancy.

Management Consider changing medication (especially if taking ACE inhibitor, ARB, or chlorothiazide) to antihypertensive drugs known to be safe in pregnancy (e.g. methyldopa, nifedipine, or β-blocker). Monitor BP carefully aiming to keep BP <150/100mmHg (<140/90mmHg if target organ damage). Pre-eclampsia is ↑ ×5.

Pregnancy-induced hypertension (PIH) ↑ BP appearing >20wk into pregnancy and resolving <3mo after delivery. Affects 10% of pregnancies and risk of pre-eclampsia is ↑. Treatment is the same as for chronic hypertension. ↑ risk of developing hypertension later in life.

Pre-eclampsia (PET)[N] Affects 5–7% of primigravida and 2–3% of all pregnancies. Multisystem disease of unknown cause, developing ≥20wk into pregnancy and resolving <10d after the birth. Untreated, may progress to eclampsia—➔ p. 1084.

Risk factors for pre-eclampsia Evaluate at booking:

Moderate risk:
- First pregnancy
- Age ≥40y
- Pregnancy interval >10y
- BMI ≥35kg/m^2 at first visit
- Family history of pre-eclampsia
- Multiple pregnancy

High risk:
- Chronic hypertension
- ↑ BP during previous pregnancy
- Chronic kidney disease
- Type 1 or type 2 DM
- Autoimmune disease, e.g. SLE or antiphospholipid syndrome

If ≥1 high risk factor or ≥2 moderate risk factors, advise aspirin 75mg from 12wk to birth and refer <20wk for specialist obstetric care.

Criteria for diagnosis
- **BP >140/90mmHg or >+30/+15mmHg from booking** The earlier in pregnancy the BP rises, the more likely the pre-eclampsia will be severe
- **Proteinuria ≥0.3g/24h** Urine dipstick is a useful screening tool—if ≥1+ protein, then probably significant—but ~25% false +ve rate

Interval for routine BP checks Pre-eclampsia is asymptomatic until its terminal phase, and onset may be rapid. Frequent BP screening is essential. Whenever you check BP, always check urine for protein.
- If no risk factors for pre-eclampsia—routine antenatal care
- If 1 moderate risk factor for pre-eclampsia, recheck BP at least every 3wk from 24–32wk gestation and at least every 2wk after 32wk gestation
- If one or more high risk factor or ≥2 moderate risk factors, monitor as directed by the specialist

Thresholds for further action Table 22.5

Risk of recurrence In subsequent pregnancy with the same partner, risk of gestational hypertension is 13–53%; risk of pre-eclampsia is ~ 16% (higher if severe pre-eclampsia). Greater risk of ↑ BP later in life.

Eclampsia ➔ p. 1084 **HELLP syndrome** ➔ p. 1084

Table 22.5 Hypertension and pre-eclampsia: thresholds for further action in the community

Clinical scenario	Clinical findings	Action
New hypertension *without* proteinuria >20wk gestation	Diastolic BP ≥90 and <100mmHg	Refer for specialist assessment[a] in <48h
	Diastolic BP ≥90 and <100mmHg with significant symptoms (below)	Refer for same-day specialist assessment[a]
	Diastolic BP ≥100mmHg	
	Systolic BP ≥160mmHg	
New hypertension *with* proteinuria >20wk gestation	Diastolic BP ≥90mmHg and new proteinuria ≥1+ on dipstick	
	Diastolic BP ≥90mmHg and new proteinuria ≥1+ on dipstick and significant symptoms (below)	Immediate admission
	Diastolic BP ≥110mmHg and new proteinuria ≥1+ on dipstick	
	Systolic BP ≥160mmHg and new proteinuria ≥1+ on dipstick	
New proteinuria *without* hypertension >20wk gestation	1+ on dipstick	Repeat pre-eclampsia assessment in <1wk
	2+ on dipstick	Refer for specialist assessment[a] in <48h
	≥1+ on dipstick and significant symptoms (below)	Refer for same-day specialist assessment[a]
Maternal symptoms *or* fetal signs/symptoms *without* new hypertension or proteinuria	Headache and/or visual disturbance with diastolic BP <90mmHg and trace or no proteinuria	Investigate cause of headache. ↓ interval to next pre-eclampsia assessment
	Epigastric pain with diastolic BP <90mmHg and trace/no proteinuria	If simple antacids are ineffective, refer for same-day specialist assessment[a]
	↓ fetal movements or small-for-gestational age infant with diastolic BP <90mmHg and trace/no proteinuria	Refer for investigation of fetal compromise. ↓ interval to next pre-eclampsia assessment

⚠ Significant symptoms indicating possible pre-eclampsia

- Epigastric pain
- Vomiting
- Headache
- Visual disturbance
- ↓ fetal movements
- Small for gestational age fetus

[a] Most obstetric departments have a day case 'step-up' assessment unit.

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Further information

Action on Pre-eclampsia (APEC) (2012) PRECOG: the pre-eclampsia community guideline. 🖱 https://action-on-pre-eclampsia.org.uk/wp-content/uploads/2012/07/PRECOG-Community-Guideline.pdf

NICE (2019) Hypertension in pregnancy: diagnosis and management. 🖱 https://www.nice.org.uk/guidance/ng133

Diabetes and epilepsy in pregnancy

Pre-existing DM Affects 2–3/1000 pregnancies. 95% have type 1 DM.

Effects on the baby

- *In utero* Large for dates or IUGR; fetal hyperinsulinaemia; ↑ congenital abnormalities (cardiac, renal, and neural tube defects); hypoxia and intrauterine death (especially >36wk)
- Postnatally Hypoglycaemia; transient tachypnoea of the newborn or respiratory distress syndrome; neonatal jaundice

Effects on the mother Problems are more common if poor control.

- In pregnancy First-trimester miscarriage; premature labour; pre-eclampsia; pyelonephritis; polyhydramnios; ↑ retinopathy
- In labour Fetal distress; obstruction (especially shoulder dystocia)

Management pre-pregnancy Suggest counselling via a diabetic specialist. Pay careful attention to diabetic control (aim blood glucose level 4–6mmol/L pre-meals). Advise folate supplements 5mg od until 13wk. Stop drugs contraindicated in pregnancy, e.g. ACE inhibitors, sulfonylureas.

Management during pregnancy Refer to an obstetrician early. Most women with type 1 DM continue to use their pre-pregnancy insulin regimen but requirements ↑ 2–3× in pregnancy. Metformin is safe in pregnancy for women with type 2 DM. USS is used to monitor fetal growth and exclude structural abnormalities. Delivery should always take place in a specialist unit with neonatal care facilities.

Retinal screening Retinopathy can worsen in pregnancy. Retinal screening is important:

- At booking For all women with pre-existing DM
- At 16–20wk If any retinopathy at booking
- At 28wk For all women with pre-existing DM (not gestational DM)

Management postnatally ↓ insulin to pre-pregnancy levels (if breastfeeding may need less). Take specialist advice regarding oral hypoglycaemics and breastfeeding.

Gestational diabetes DM with onset/first recognition in pregnancy. Affects 2% of pregnancies and usually develops in the second trimester. Lower risk of congenital malformation than if pre-existing DM. Intensive management can achieve almost normal rates of macrosomia and neonatal hypoglycaemia, but there is debate whether that is necessary (💣). *Screening*: ➔ p. 786

Risk factors

- Previous macrosomic baby (≥4.5kg)
- Polycystic ovarian syndrome (PCOS)
- BMI >30kg/m^2
- Previous gestational DM
- DM in a first-degree relative
- Family origin associated with high prevalence of DM: South Asian (especially Indian subcontinent); black Caribbean; Middle Eastern

Management Initially diet; some may require metformin (unlicensed) and up to 30% require insulin. Insulin is stopped immediately postpartum. Check a 6wk postpartum oral glucose tolerance test. Gestational DM usually recurs in future pregnancies; >30% develop DM in <10y.

Glycosuria in pregnancy Do not routinely screen for glycosuria in pregnancy. Pregnant women have ↓ renal threshold and a physiologically ↑ plasma glucose, so dipstick testing gives a high false +ve rate. If glycosuria is detected, repeat—if still +ve, arrange an oral glucose tolerance test.

Epilepsy in pregnancy 90% have normal pregnancies/healthy babies.

In utero effects on the fetus Many antiepileptic drugs are teratogenic—especially if taken in the first trimester and if taking ≥2 drugs. Sodium valproate is associated with the highest risk of congenital malformation (1 in 10) and developmental delay (4 in 10). Topiramate is associated with ↑ risk of cleft palate if taken in the first trimester.

Peri/postnatal effects on the baby Haemorrhagic disease of the newborn is associated with antiepileptic drugs—all babies should have IM vitamin K at birth; the child has ↑ risk of epilepsy.

Effects on the mother

- In pregnancy 10% have ↑ fit frequency. Risk to the fetus from most fits is low; status epilepticus is associated with high infant and maternal mortality
- In labour/puerperium 2–6% have a fit during labour or <48h after delivery

Management pre-pregnancy Discuss risks of pregnancy and epilepsy/antiepileptic medication with all epileptic women of childbearing age—whether or not contemplating pregnancy. Suggest referral to neurology for optimization of antiepileptic regimens prior to pregnancy. Advise folic acid 5mg od from when pregnancy is planned until 13wk gestation.

> ⚠ Avoid prescribing sodium valproate to women of childbearing age. If essential, ensure a pregnancy prevention plan is in place and that the woman is informed about potential teratogenic effects both verbally and in writing.

Management during pregnancy Refer for specialist obstetric care—joint management by an obstetrician and neurologist is ideal. Antiepileptic dose adjustments may be needed. Delivery should occur in a specialist centre where fits can be managed. Encourage women to notify the UK Epilepsy and Pregnancy Register (🖱 www.epilepsyandpregnancy.co.uk) of their pregnancy.

Management postnatally Breastfeeding is not contraindicated if taking monotherapy, but infants should be monitored for side effects of antiepileptic medication. If drug dose is ↑ in pregnancy, it may need to ↓ after delivery. All babies should have IM vitamin K due to ↑ risk of haemorrhage. Risk of injury to the child from maternal seizure is low. Discuss child care and minimizing risks to the child from the mother's epilepsy.

Further information

MHRA (2018) Valproate use by women and girls. 🖱 www.gov.uk/guidance/valproate-use-by-women-and-girls

NICE (2015) Diabetes in pregnancy: management from preconception to the postnatal period. 🖱 www.nice.org.uk/guidance/ng3

NICE (2012, updated 2018) The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care. 🖱 www.nice.org.uk/guidance/cg137

Intrauterine growth, multiple pregnancy, and malpresentation

Intrauterine growth restriction (IUGR)^{G} Babies may be small because they are premature, small for their gestation, or a combination of the two. Babies weighing <10th centile weight for their gestational age (IUGR) have different problems to premature babies.

Predisposing factors The major antenatal indicator for IUGR is low maternal weight at booking (<51kg). *Others include*:

- Multiple pregnancy
- Malformation
- Infection
- Maternal smoking
- Maternal DM
- Pre-eclampsia
- Severe maternal anaemia
- Maternal heart or renal disease
- Previous history of small baby
- Low weekly maternal weight ↑ (<0.2kg)

Antenatal detection Difficult to detect; about half are not detected until after birth. Most GPs will encounter IUGR when the community midwife does a routine antenatal check and finds the symphysis–fundal height (SFH) is less than would be expected for the gestation. Other suspicious signs are oligohydramnios and poor fetal movements. Confirm suspicions with USS then seek specialist obstetric advice. Where the head circumference is relatively spared, suspect placental insufficiency.

Consequences

- **Labour** More susceptible to hypoxia in labour, so require monitoring in a specialist unit where Caesarean section facilities are available and there is paediatric back-up
- **Postnatal problems** Susceptible to neonatal hypoglycaemia and jaundice. Babies <2kg may have problems with temperature regulation and require incubator facilities
- **Long-term effects** More prone in later life to cardiovascular disease and type 2 DM

Oligohydramnios Liquor volume <500mL. Rare. Associated with:

- Prolonged pregnancy
- PROM (➔ p. 812)
- Placental insufficiency
- Fetal abnormality (renal agenesis, urethral aplasia)

Confirm diagnosis with USS then refer for specialist obstetric assessment.

Large for dates Consider:

- Multiple pregnancy
- Maternal DM
- Fetal abnormality
- Polyhydramnios
- Molar pregnancy
- Large baby (>90th centile)—may have past history of large babies

Refer for USS to confirm diagnosis and exclude fetal abnormality or multiple pregnancy. Check maternal oral glucose tolerance test.

Polyhydramnios Liquor volume >2L; affects 0.15% pregnancies. *Causes*:

- **Fetal abnormality** e.g. hydrops fetalis; anencephaly (no swallowing reflex); spina bifida; oesophageal or duodenal atresia; umbilical hernia; ectopia vesicae (ectopic bladder)

- **Maternal** e.g. DM, multiple pregnancy
- **No cause found** (30–50%)

Risks Premature labour; malpresentation; cord prolapse; placental abruption; PPH.

Management Refer for USS to confirm diagnosis. Check maternal oral glucose tolerance test and refer for specialist obstetric advice.

Multiple pregnancy Detected on early antenatal USS. *Incidence*: twins—1 in 105 (1 in 3 identical); triplets—1 in 10,000. *Predisposing factors*:

- Previous twins
- Family history of non-identical twins
- Race: most common among African black ♀; least common in Japanese ♀
- ↑ with maternal age
- Infertility treatment—induced ovulation (e.g. clomifene); IVF and other assisted reproduction techniques

Management Refer for specialist obstetric care. Monochorionic twins are significantly higher risk than dichorionic twins. *Complications*:

- **In pregnancy** Hyperemesis; anaemia; polyhydramnios; pre-eclampsia (↑ ×3); APH; placenta praevia; placental abruption
- **In labour** Malpresentation; cord prolapse; fetal distress (↑ Caesarean section rate); PPH
- **Fetus** ↑ perinatal mortality (×5); prematurity; IUGR; malformations (↑ ×2–4); twin–twin transfusion may result in 1 twin being plethoric (and jaundiced later) and the other anaemic

Breech babies[G] 3–4% babies at term. Higher incidence <37wk. Associated with ↑ risk of cerebral palsy as breech presentation is more common in premature infants and those with congenital malformation. *Risk factors*: bicornuate uterus; fibroids; placenta praevia; oligohydramnios.

Management The baby may turn spontaneously—especially if <36wk gestation. If a baby is found to be breech at ≥36wk gestation, confirm breech position and position of the placenta on USS and refer for specialist obstetric advice. *Specialist options*: external cephalic version (ECV); vaginal breech delivery; elective Caesarean section.

> ⚠ 10–15% of breech positions are discovered for the first time late in labour. If delivering at home or in a community unit, arrange transfer to a specialist unit immediately.

Follow-up Congenital hip problems are more common in breech babies—refer all breech babies routinely for hip USS even if examination in the first 24h is normal.

Further information

RCOG (2013) Small-for-gestational-age fetus: investigation and management. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg31

RCOG (2017) Management of breech presentation. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg20b

Information and support for multiple pregnancy

Twins and Multiple Births Association (TAMBA) 🖱 www.tamba.org.uk

Ante- and postpartum haemorrhage

Bleeding in early pregnancy ➔ p. 766.

Antepartum haemorrhage Any bleeding in pregnancy >24wk gestation (or the point of fetal viability) is an antepartum haemorrhage (APH).

Causes

Uterine:

- Abruption (➔ p. 1084)
- Placenta praevia
- Vasa praevia
- Circumvallate placenta
- Placental sinuses

Lower genital tract:

- Cervical—polyp; erosion; carcinoma; cervicitis
- Vaginitis
- Vulval varicosities

⚠ *Action* ALWAYS admit to a specialist obstetric unit. If bleeding is severe, admit via an emergency ambulance, and while awaiting transport, raise legs; give O_2 via face mask; if possible, gain IV access, take blood for FBC and cross-matching, and start IV infusion. NEVER do a vaginal examination—placenta praevia bleeds +++.

Placenta praevia^{G} Occurs when the placenta lies within the lower uterine segment. *Incidence*: 1 in 4 routine anomaly scans done at 19wk gestation show a low-lying placenta—5% stay low at 32wk; <2% at term.

Associations

- ↑ with parity
- Age >35y
- Smoking
- Twins
- Preterm delivery
- Previous Caesarean section
- Endometrial damage (e.g. history of dilatation and curettage, TOP)
- Placental pathology (marginal/velamentous cord insertions, succenturiate lobes, bipartite placenta)
- Previous placenta praevia (recurrence rate 4–8%)

Management If the placenta covers/overlaps the cervical os at the 18–20wk routine USS, a follow-up USS is performed at 32wk to determine whether the placenta is moving out of the lower segment. If the placenta remains low, management depends on whether the placenta covers the internal os (major placenta praevia) or not (minor placenta praevia). Major placenta praevia always requires delivery by Caesarean section. Normal delivery in a specialist unit may be attempted with minor placenta praevia if the placental edge is >2cm from the internal os.

❗ Women who have anterior placenta praevia on routine anomaly scanning and who have had a previous Caesarean section may have placenta accreta.

Maternal complications

- APH—typically painless bleeding >20wk with a peak incidence at 34wk
- Malpresentation—35% breech presentation or transverse lie
- Placental problems—placenta accreta and percreta especially with a history of previous Caesarean section; abruption
- Postpartum haemorrhage

Fetal complications IUGR (15%); premature delivery; death.

Primary postpartum haemorrhage (PPH) Loss of >500mL blood within 24h of delivery. Affects 1 in 100 deliveries. May occur in the community after home delivery, delivery in a community obstetric unit, or after rapid discharge from a consultant-led unit.

Causes The four Ts:

- Tone—uterine atony (90%)
- Tissue—retained products of conception
- Trauma—of the genital tract
- Thrombin—clotting disorders

Risk factors

- Age >40y
- Obesity (BMI >35kg/m^2)
- Pre-eclampsia or gestational hypertension
- Large placental site (e.g. multiple pregnancy or baby >4kg)
- Abruption—known or suspected
- Prolonged labour (>12h)
- Asian ethnicity
- Pyrexia in labour
- Low placenta
- Past history of PPH
- Anaemia (Hb <9g/dL)
- Caesarean section delivery (emergency or elective)
- Operative vaginal delivery and/or mediolateral episiotomy
- Retained placenta (➔ p. 1085)

⚠ *Action* Call emergency ambulance for immediate transfer to hospital.

- If possible, gain IV access, take blood for FBC/cross-match, start IV infusion, and give ergometrine 0.5mg slowly IV (if available)
- Give high-flow O_2 via face mask as soon as possible
- If placenta is not delivered, try to deliver it by controlled cord traction
- Check for trauma; apply pressure/repair any visible bleeding point. Bimanual pressure on the uterus (rubbing up the fundus) may ↓ immediate loss
- Some community units keep carboprost 250 micrograms for emergency use. Use if available—1mL by deep IM injection—repeat after 15min. ❗ Contraindicated in women with asthma

Secondary PPH Excessive blood loss PV >24h after delivery. *Peak incidence*: 5–12d after delivery.

Cause Postpartum infection—sometimes associated with retained placental tissue or clot.

⚠ *Action*

- If the woman is unwell (shocked or toxic) admit to an obstetric unit for further investigation, IV antibiotics ± evacuation of retained products of conception
- If bleeding is slight, manage conservatively. Take a vaginal swab and start oral antibiotics—amoxicillin 500mg tds and metronidazole 400mg tds. Consider referral for USS and/or obstetric review if not settling

Further information

RCOG (2011, reviewed 2014) Antepartum haemorrhage. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg63

RCOG (2016) Postpartum haemorrhage. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg52

RCOG (2018) Placenta praevia and placenta accreta: diagnosis and management. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg27a

Labour

47% of deliveries are 'normal', i.e. occur without surgical intervention, use of instruments, induction, epidural, or general anaesthetic. Intrapartum care can be provided by GPs in the UK as a National Enhanced Service.

Braxton Hicks contractions Irregular tightenings of the uterus. Start ≥30wk gestation (common >36wk). May be uncomfortable but not painful.

Premature rupture of membranes (PROM) Rupture of membranes before labour starts. Usually presents with a gush of clear fluid (± an audible pop) followed by uncontrolled leakage. If chorioamnionitis is present the woman may have abdominal pain and feel unwell. Difficult to distinguish clinically from profuse vaginal discharge or incontinence of urine. Check temperature, pulse, and BP and do a routine obstetric examination (including fetal heart). ❗ Do not perform a vaginal examination as repeated examinations can introduce infection.

Management

- Evidence of infection Admit for specialist obstetric care
- <37wk gestation and suspected PROM Admit to specialist obstetric unit for further assessment
- ≥37wk gestation If no signs of spontaneous labour admit for specialist obstetric assessment within 24h

Premature labour Any labour <37wk gestation. *Prevalence*: 6%—1 in 4 elective due to maternal/fetal problems. Largest contributor to neonatal morbidity/mortality in industrialized countries.

Causes of spontaneous premature labour Unknown (40%).

- Cervical incompetence
- Multiple pregnancy
- Uterine abnormality
- DM
- Pyelonephritis or other sexually transmitted or urinary infection
- Polyhydramnios
- APH

Presentation Premature rupture of membranes or contractions. If suspected admit immediately to obstetrics for further assessment.

Prolonged pregnancy/post-maturity Due date is traditionally based on pregnancy lasting 40wk or 280d from the date of the LMP. Now most women have an early dating USS to confirm their due date. However, it is normal to deliver any time from 37 to 42wk. At 40wk gestation, 65% will spontaneously go into labour in the next week but 15% of women have not gone into labour by 42wk. Perinatal mortality rate is ↑ ×2 from 42–43wk and ↑ ×3 >43wk, so induction of labour is indicated if a pregnancy lasts >42wk.

Initial management Membrane sweep (usually performed by midwife). If that is ineffective, refer for formal induction of labour (➔ p. 814). If referral is declined, ↑ antenatal monitoring to 2× weekly cardiotocography and USS (to measure maximum amniotic pool depth) as markers of fetal well-being.

Normal labour Occurs ≥37wk gestation and results in vaginal delivery of a baby in <24h. Often heralded by a 'show' consisting of mucus ± blood and/or spontaneous rupture of membranes ('waters going').

- 1st stage of labour Time from the onset of regular contractions until the cervix is fully dilated
- 2nd stage of labour Time from complete cervical dilatation until the baby is born. The mother has a desire to push
- 3rd stage of labour Delivery of the placenta

Pain relief for labour Most women experience pain in labour. Strategies for pain relief include:

- Self-help Keep fit in pregnancy, relaxation techniques, breathing exercises, warm bath/birthing pool
- TENS Machines are available to hire from most obstetric units and some retail outlets
- Entonox® Takes 30–45s to have effect—advise women to start inhaling it as soon as the contraction starts
- Injected opioids e.g. pethidine
- Epidural
- Pudendal block Used for instrumental delivery

Advise women to discuss options with their midwife. Antenatal classes dealing with pain relief significantly ↑ women's confidence in managing labour pains.

Epidural Effective method of analgesia available in most hospital units. Initiated once in established labour (cervix >3cm dilated). Regular BP, pulse, and fetal heart monitoring is required. *Particular indications*:

- Occipito-posterior (OP) position
- Breech
- Multiple pregnancy
- Pre-eclampsia
- Forceps delivery
- Maternal medical conditions, e.g. cardiac

Epidural complications during labour

- Postural hypotension
- Urinary retention
- ↑ need for instrumental delivery due to pelvic floor muscle paralysis

Epidural complications post-delivery Urinary retention, headache (especially if dural puncture).

Meconium-stained liquor Passage of fresh meconium (dark green, sticky, and lumpy) during labour may be a sign of fetal distress. Transfer immediately to a consultant unit for further evaluation.

Management Paediatrician should be present at delivery. Do not perform oropharyngeal suction if there is no evidence of fetal hypoxia.

Dystocia Difficulty in labour. May be due to problems relating to the baby, birth passage, or action of the uterus. Neonatal mortality and maternal morbidity both ↑ with duration of labour. *Possible causes*:

- Pelvic abnormality
- Shoulder dystocia (➔ p. 1085)
- Abnormal presentation
- Uterine dysfunction
- Cervical dystocia
- Cephalo-pelvic disproportion

Management If a patient in labour at home or in a community unit fails to progress as expected, admit immediately to a specialist unit for consideration of intervention to speed the labour or Caesarean section. Shoulder dystocia is an obstetric emergency—➔ p. 1085.

Induction of labour$^{\mathrm{N}}$ Performed when it is felt the baby is better off out than in (~20% of deliveries). Only undertaken in units with facilities for continuous fetal monitoring and emergency Caesarean section.

Procedure involves Assessment of the cervix; vaginal prostaglandins; 'sweeping' of the membranes; artificial rupture of the membranes and/or IV oxytocin to maintain contractions.

Reasons for induction of labour include
- Post-maturity (most common)—offered from 41–42wk
- Premature rupture of membranes
- Intrauterine death

Assisted delivery$^{\mathrm{G}}$ (Table 22.6) Forceps and ventouse are used in ~11% of deliveries in the UK (range 4–25% between hospitals). Assisted delivery should only be performed with adequate analgesia (usually epidural or pudendal block) and by experienced practitioners.

Caesarean section (LSCS)$^{\mathrm{N}}$ (Table 22.7) Rate in England and Wales is 24.8% (range 10–65% between different hospitals). 10% are elective and usually performed at >39wk to minimize risk of respiratory complications; the other 11–12% occur after labour has started. Regional anaesthesia for LSCS is safer for mother and child than a GA.

Reasons for emergency LSCS Failure to progress (25%); presumed fetal compromise (28%); breech (14%).

Planned LSCS is indicated for
- Breech (where external cephalic version has failed)
- Multiple pregnancies where the first twin is not cephalic
- Placenta praevia or placenta accreta
- Some HIV-positive women and those with primary HSV in the third trimester to ↓ virus transmission

❗ Maternal request may be an indication for LSCS; GPs should discuss risks and benefits if a request is made, and if the patient still requests a LSCS, refer for a consultant opinion.

Further information

NICE (2008) Inducing labour. 🖱 www.nice.org.uk/guidance/cg70
NICE (2011, updated 2019) Caesarean section. 🖱 www.nice.org.uk/guidance/cg132
NICE (2014, updated 2017) Intrapartum care for healthy women and babies. 🖱 www.nice.org.uk/guidance/cg190
NICE (2015, updated 2019) Preterm labour and birth. 🖱 www.nice.org.uk/guidance/ng25
RCOG (2011, reviewed 2014) Operative vaginal delivery. 🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg26

Information for women

Emma's diary Labour information pack. 🖱 www.emmasdiary.co.uk/about-us/our-publications/labour-information-pack
National Childbirth Trust (NCT) ☎ 0300 330 0700 🖱 www.nct.org.uk

Table 22.6 Forceps and ventouse

	Forceps	Ventouse
Indication	Delayed 2nd stage of labour	Delayed 2nd stage of labour
Procedure	● Wrigleys' forceps—for 'lift-out' deliveries ● 'Neville Barnes' forceps—for high deliveries ● 'Keilland's' forceps—if rotation is required	● The vacuum extraction cup is applied to the baby's head, suction is applied, and traction aids delivery ● Ventouse allows rotation if the baby is malpositioned
Early complications	● Maternal trauma (episiotomy always needed) ● Fetal facial bruising ● Fetal facial nerve paralysis	● 'Chignon' develops on the baby's head—resolves in ≤2d ● Cephalohaematoma ● Retinal haemorrhage ● Neonatal jaundice (but no ↑ need for phototherapy)
Longer-term complications	↑ risk of maternal faecal incontinence	
Comparison	Ventouse has ↑ failure rate compared to forceps but no ↑ LSCS rate ↓ requirement for regional anaesthesia with ventouse deliveries compared to forceps deliveries Forceps result in more maternal trauma than ventouse deliveries	

Table 22.7 Comparison of Caesarean section and vaginal birth

	Complications
↑ with LSCS	Mother—this pregnancy Abdominal pain; bladder or ureteric injury; hysterectomy; maternal death; need for further surgery; need for admission to intensive care/high dependency unit; thromboembolism; length of hospital stay; need for readmission Mother—future pregnancies Not having more children; antepartum stillbirth; placenta praevia; uterine rupture Baby Neonatal respiratory problems
No difference	Mother Haemorrhage; infection; genital tract injury; faecal incontinence; back pain; dyspareunia; postnatal depression Baby Death (except breech); intracranial haemorrhage; brachial plexus injuries; cerebral palsy
↓ with LSCS	Mother Perineal pain; urinary incontinence; uterovaginal prolapse

Maternal postnatal care

Postnatal care, from hospital discharge until 14d after delivery, excluding the neonatal check, is provided as an Additional Service (Maternity Services). Payment is included in the Global Sum ('opting out' results in a 2.1% ↓).

Puerperium 6wk period immediately after delivery. Initial checks—Box 22.3. Most women in the UK spend ≥6h after delivery in hospital. After discharge, the midwife continues to visit for 2wk after the birth and then the health visitor takes over. GPs often only see women in the puerperium to do the routine 6wk postnatal check. Arrange additional reviews as needed.

Mother's 6wk postnatal check Discuss:

- Any problems in pregnancy or delivery
- Current problems the mother has—specifically enquire about persistent vaginal loss, bladder/bowel control, and any sex-related problems
- Problems with the baby—including feeding and worries about hearing/vision
- Contraception

Examination/investigation

- BP and weight—if overweight discuss weight control—➲ p. 152
- Examination—uterus should not be palpable per abdomen; only do vaginal examination if problem with tear/episiotomy, persistent vaginal bleeding, or pain. Delay overdue cervical smear until >12wk after delivery
- Screen for depression—4–6wk and 3–4mo postnatally (➲ p. 819)
- Check Hb if anaemic postnatally
- Arrange for rubella immunization if not immune antenatally—ensure effective contraception for 3mo. Check immunity 3mo after vaccination. Reassure safe to breastfeed after immunization

The baby's 6wk developmental check ➲ p. 828

Contraception after pregnancy[G] ❗ Contraception is not needed until 21d after childbirth. Interpregnancy interval of <12mo is associated with ↑ risk of pre-term birth, low birthweight, and small-for-gestational age babies.

Emergency contraception Indicated if unprotected sexual intercourse ≥21d after childbirth. Oral levonorgestrel 1.5mg and ulipristal acetate (UPA) 30mg can be used from 21d; Cu-IUD can be used from 28d. If UPA emergency contraception, should not breastfeed and express/discard milk for 1wk.

Combined hormonal contraception (CHC) If not breastfeeding and no risk factors for venous thromboembolism (VTE), start CHC ≥21d after delivery. Do not use CHC for ≥6wk if any of the following:

- Immobility
- BMI ≥30kg/m^2
- Smoking
- Transfusion at delivery
- Postpartum haemorrhage
- Caesarean delivery
- Pre-eclampsia

Progestogen-only contraception (IUS, POP, injectable or implant) Start at any time after childbirth, including immediately after delivery. No evidence of any adverse effects on lactation or infant growth/development.

Intrauterine contraception Can be inserted immediately after birth or <48h after uncomplicated Caesarean section/vaginal birth. After 48h, insertion should be delayed until 28d after childbirth.

Box 22.3 Standard early postnatal checks

- **Rhesus status** If the mother is RhD –ve and the baby RhD +ve, ensure anti-D is given <72h after delivery—➔ p. 770
- **Hb on day 5 after delivery** (after the postpartum diuresis). If Hb is <10g/dL, continue iron supplements for 3mo
- **Temperature, pulse, and BP** ↑ BP is associated with pre-eclampsia. It usually resolves <48h after delivery. Fever may indicate infection
- **Fundus** Day 1 = 24wk gestation size (up to umbilicus); day 5 = 16wk gestation size; by day 10, the uterus should not be palpable per abdomen. Persistent bulkiness suggests retained products of conception—refer for USS
- **Pain** Breast, abdominal, perineal, legs
- **Vaginal loss** Red, then brown, then yellowish over the first week, then serous for 3–6wk. Any fresh, red bleeding is abnormal after the initial 2–3d
- **Moving about** Women should try to get mobile as soon as possible after delivery to ↓ the risk of DVT
- **Feeding the baby** ➔ p. 848
- **Mental state**

Other methods

- **Sterilization** A safe option for contraception after pregnancy. If having an elective Caesarean section, the option of sterilization should be discussed and agreed >2wk before the procedure
- **Male and female condoms** Can be safely used by women after childbirth
- **Diaphragm** Use other methods until >6wk after childbirth then re-fit the diaphragm; size required may change after childbirth
- **Fertility awareness methods** Can be used but signs/symptoms of fertility can be difficult to interpret after childbirth/during breastfeeding

Breastfeeding and contraception If <6mo postpartum, amenorrhoeic, and fully breastfeeding, there is low chance of pregnancy without contraception (~2/1000 women). If supplementary bottle feeding, baby is weaned, or vaginal bleeding (except occasional spotting), assume the woman is fertile. Barrier, intrauterine methods, and barrier methods are safe if breastfeeding. Wait >6wk after delivery before starting CHC for breastfeeding women.

Additional contraception (e.g. barrier method/abstinence) Is required if hormonal/intrauterine contraception is started ≥21d after childbirth:

- **POP** 2d if not started in days 1–5 of the cycle or cycle not re-established
- **Injectable/implant** If given for the first time after day 5 of the cycle (or cycle not re-established), check not pregnant, give the first injection, and advise to use an additional form of contraception for 7d
- **IUS/Cu-IUD** If not inserted immediately after birth, and after day 7 of the cycle (or cycle not re-established), use additional contraception for 7d
- **CHC** 7d if not started in days 1–5 of the cycle or cycle not re-established (9d if not started on day 1 of the cycle and estradiol valerate/dienogest pill)

Further information

FSRH (2017) Contraception after pregnancy. 🔗 www.fsrh.org/documents/contraception-after-pregnancy-guideline-january-2017

NICE (2006, updated 2015) Postnatal care up to 8 weeks after birth. 🔗 www.nice.org.uk/guidance/cg37

NICE (2014, updated 2018) Antenatal and postnatal mental health: clinical management and service guidance. 🔗 www.nice.org.uk/guidance/cg192

Common postnatal problems

Abdominal pain Cramping, 'period like' for the first 1–2wk after delivery, especially when breastfeeding, due to the uterus contracting down or involuting. Suspect infection if offensive lochia, fever, the uterus stops getting smaller day by day, or is still palpable per abdomen 10d after delivery.

Breast soreness The breasts become engorged ('the milk comes in') 3–5d after the birth and may be quite painful. Support with a well-fitting maternity bra, day and night. Express milk if still painful—a warm bath may help. *Other problems*:

Sore/cracked nipples Try topical remedies and/or nipple shields. Consider advice from a breastfeeding advisor—may be a positioning problem.

Skin infection Localized soreness, pain around the areola ± nipple, or in the breast after a feed—usually due to candida infection. Treat mother and baby with miconazole oral gel.

❗ Severe knife-like pain in breast during and for up to 1h after feeding suggests deeper infection—treat mother additionally with fluconazole 150mg stat and then 50mg bd for 10d (unlicensed use). Symptoms usually resolve in <3d.

Blocked duct Hard, tender lump in the breast. Advise the mother to massage that area of the breast while feeding or expressing milk.

Mastitis Tender, hot, reddened area of breast ± fever. Treat with flucloxacillin 500mg qds and NSAID, e.g. ibuprofen 400mg tds prn. Continue breastfeeding or express milk to prevent milk stagnation if too painful for feeding.

Breast abscess Admit for incision and drainage.

Dyspareunia following perineal trauma. Almost always settles without need for surgery.

Fever See puerperal pyrexia—➔ p. 820.

Hair loss Hair becomes thicker in pregnancy and these hairs are all shed at about the same time ~5–6mo postpartum. Reassure. Hair loss reverts to normal levels within 2–3mo. If severe, persistent, or accompanied by tiredness, consider hypothyroidism (➔ p. 335)—check TFTs.

Haemorrhoids Common and painful. *Try*:

- Local ice packs (frozen fingers of rubber gloves are the right shape)
- Topical preparations, e.g. Proctosedyl®
- Resting lying on one side
- Keeping stools soft using a stool softener
- Advising women to wash the haemorrhoids with cool water after opening bowels and gently push them through the anus (if possible)

Persistent lochia Bleeding (lochia) >6wk postpartum. *Causes*:

- Infection
- Retained products of conception
- Unhealed tears—cervical, vaginal, or perineal
- Resumption of normal cycle
- Side effects of contraception (e.g. POP, depot injection)
- Other cervical or uterine pathology

Management

- Examine uterus per abdomen and do a bimanual vaginal examination to check involution. Perform a speculum examination and send a vaginal swab for M,C&S
- If offensive loss or systemic symptoms/signs of infection, treat with antibiotics as for endometritis (➔ p. 820). Otherwise, arrange USS
- If not settling and no cause is found, refer to gynaecology

Mood disturbance

Baby blues Very common—women become tearful and low within the first 10d of delivery. Be supportive. Usually resolves.

Postnatal depression Common (10–15% mothers) reaching a peak ~12wk after delivery—although symptoms are almost always present at 6wk.

Screening for postnatal depression Often mothers do not report symptoms. NICE recommends screening all mothers for depression 4–6wk and 3–4mo postnatally by asking:

- During the past month, have you often been bothered by feeling down, depressed, or hopeless?
- During the past month, have you often been bothered by having little interest or pleasure in doing things?

If the woman answers 'yes' to either of the initial questions, ask: 'Is this something you feel you need or want help with?'

Risk factors for postnatal depression Include:

- Depression during pregnancy
- A bad birth experience
- Social problems (e.g. poor social support, financial problems)
- Past medical history or family history of depression or postnatal depression
- Alcohol or drug abuse

Management of postnatal depression

- Talk through the problems. Refer to health visitor for support
- Give information, e.g. self-help groups, mother-and-baby groups
- Consider checking TFTs—especially if presenting with tiredness
- Consider referral for psychological therapies such as CBT
- Consider antidepressant medication. Of the SSRIs, sertraline 50mg od is the safest. In all cases, monitor the baby for unwanted side effects (e.g. drowsiness, respiratory depression). If not breastfeeding, fluoxetine 20mg od is the most effective antidepressant in trials
- Monitor progress using depression questionnaires, e.g. Edinburgh Postnatal Depression Scale

Puerperal psychosis Much rarer than postnatal depression (1 in 500 births). Suspect if severe depression; high suicidal drive; mania; psychotic symptoms. In all cases seek expert help from a psychiatrist. Consider admission—under a Section if necessary. Risk of recurrence is 20%—but 50% will never be mentally ill again.

⚠ Refer to the perinatal mental health team immediately if any risk of self-harm, suicide, or harm to the baby.

Perineal bruising Can be very painful. Advise regular analgesia, e.g. paracetamol 1g qds ± ibuprofen 400mg tds, ice packs. Ultrasound can help—consider referral to physiotherapy.

Poor abdominal and pelvic muscle tone Classes for postnatal exercise to re-tone the body are available both on dry land and in the swimming pool at most leisure centres. Pelvic floor exercises can be started <1d after delivery (Box 22.4). Good leaflets explaining these are available from physiotherapists, local maternity units, and the NCT.

Puerperal pyrexia Temperature >38°C within 14d of delivery or miscarriage. 90% infections are in the urinary or genital tracts. Ask about:

- Urinary symptoms
- Colour and smell of lochia
- Abdominal pain
- Breast symptoms
- Any other symptoms (e.g. cough, sore throat)

Examine fully including bimanual VE and send MSU and vaginal swabs for M,C&S. *Potential obstetric causes:*

Superficial perineal infection Complicates tear or episiotomy—treat with flucloxacillin 500mg—1g qds.

Endometritis Presents with offensive lochia, lower abdominal pain, and a tender uterus. Treat with amoxicillin 500mg tds and metronidazole 400mg tds or co-amoxiclav 375mg tds. If not settling in <48h or very unwell, admit for IV antibiotics.

Mastitis See breast soreness—➔ p. 818.

DVT or PE Can present with pyrexia. Refer to exclude DVT if any leg pain and to exclude PE if chest pain/breathlessness.

Superficial thrombophlebitis Affects 1% women. Presents with a tender (usually varicose) vein. Exclude DVT. Recovery usually occurs within a few days. Meanwhile advise the woman not to stand still and, when sitting, to elevate the leg above waist height. Support the leg, e.g. with an elasticated stocking, and try applying an ice pack to the affected area. NSAIDs, e.g. ibuprofen 400mg tds prn, may help.

Tiredness Very common in the first few months after delivery but it may be the presenting feature of postnatal depression, anaemia, or hypothyroidism. Check FBC and TFTs.

Transient autoimmune thyroiditis Up to 10% women 1–3mo after delivery. Usually presents with fatigue and lethargy.

Hypothyroidism Treat with levothyroxine for 6mo then stop for 6wk and repeat TFTs. Follow-up with annual TFTs—1 in 5 go on to develop permanent hypothyroidism.

Hyperthyroidism Refer to an endocrinologist—antithyroid treatment is not normally required but symptom control may be necessary.

Box 22.4 Pelvic floor exercises—basic techniques

- **Exercise 1** Advise the woman to pull up her pelvic muscles as if stopping herself from passing urine and hold that position for a count of 10
- **Exercise 2** Advise the woman to pull up her pelvic muscles as in exercise 1, but then relax and contract them rapidly 4 times

These exercises should be repeated as many times daily as possible long term.

Further information

NICE (2006, updated 2015) Postnatal care up to 8 weeks after birth. 🖱 www.nice.org.uk/guidance/cg37
NICE (2014, updated 2018) Antenatal and postnatal mental health: clinical management and service guidance. 🖱 www.nice.org.uk/guidance/cg192

General advice and support for postnatal women

Family Planning Association 🖱 www.fpa.org.uk
National Childbirth Trust (NCT) ☎ 0300 330 0700 🖱 www.nct.org.uk

Advice and support for women with postnatal depression

Association for Postnatal Illness Support and befriending by women who have suffered postnatal depression/puerperal psychosis. ☎ 020 7386 0868 🖱 https://apni.org/
Royal College of Psychiatrists Information sheet on postnatal depression. 🖱 https://www.rcpsych.ac.uk/healthadvice/problemsanddisorders/postnataldepression.aspx

Stillbirth and neonatal death

Stillbirth is a term applied to those babies born dead after 24wk gestation. Affects 1 in 250 pregnancies in the UK. Death may occur *in utero* or during labour. Usually presents with a lack of fetal movements and on examination no fetal heart can be detected. If suspected refer as an emergency to the nearest obstetric unit for confirmation of intrauterine death by USS.

Management In hospital, mothers of babies who have died *in utero* are usually induced. Samples are routinely taken from mother and baby to try to determine cause of death.

Common causes Pre-eclampsia; IUGR; renal disease; DM; infection; malformation; post-maturity; abruption; knots in the cord. No cause is found for 1 in 5 stillbirths.

After discharge Make contact with the parents as soon as possible.

Lactation suppression Offer cabergoline 1mg as a single dose.

Registration of stillbirth Certificate of stillbirth is issued by the obstetrician. This must be taken to the Registrar of deaths <42d after the birth. Parents are issued with a certificate of burial/cremation and registration certificate to keep. The child's name may be entered on the registration certificate.

Funeral Parents have the option of a free hospital funeral. Burial is usually in an unmarked multiple occupancy grave. Parents may pay for a single occupancy grave or cremation. Alternatively, parents may pay for a private funeral.

Benefits All UK maternity benefits are payable after stillbirth—➔ p. 765.

Follow-up Routinely arranged by the specialist obstetrician to discuss reasons for the stillbirth and implications for future pregnancies. Primary care follow-up is essential. Stillbirth is a huge burden to come to terms with. Parents do not have the regular contact with medical staff a baby brings. Ensure regular follow-up by a member of the primary care team. Broach the issues brought up by the baby's death directly. Offer an open door. Give information about support organizations, e.g. SANDS. Advise waiting 6mo–1y before embarking on another pregnancy.

Neonatal death Death of an infant <28d old. Rare in the community. In the UK, all deaths of children <18y are subject to review by the local child death review panel (➔ p. 905) and should be notified to it immediately. If the death is expected, the GP will be allowed to issue a special death certificate. If unexpected, the case will be referred to the police/coroner. Offer lactation suppression with cabergoline 250 micrograms bd for 2d, and follow-up as for stillbirth.

Further information

RCOG (2010, updated 2017) Late intrauterine fetal death and stillbirth.
🖱 www.rcog.org.uk/en/guidelines-research-services/guidelines/gtg55/

Patient support and information

Stillbirth and Neonatal Death Society (SANDS) ☎ 0808 164 3332
🖱 www.uk-sands.org

‘Children are one third of our population and all our future’

US Select Panel for Promotion of Child Health (1981)

❗ In other sections of this book, where management differs from the norm for children, the text is highlighted in a box marked with this symbol.

Chapter 23

Child health

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Child health promotion

Patients ≤15y comprise 19% of the average practice list; those <5y see their GP ~6×/y; more often than any other age group, except the elderly.

The Healthy Child Programme Designed to promote good health and well-being for 0–19y-olds. It moves from rigid developmental screening to a more flexible assessment of the child within the family context. It aims to identify families in need of extra support (e.g. parental mental health issues, low income, poor living conditions) early so that extra support can be provided. It includes:

- Health promotion beginning antenatally and continuing to teenage years covering the full range of child health issues, e.g. diet, safety, substance abuse (drugs, smoking, and alcohol), teenage sexual health
- Childhood screening
- Immunization—➔ p. 619
- Health and development reviews—to monitor the child's development, the strengths/weaknesses of the family, and to discuss the parents' hopes and concerns, followed by early intervention as required

The delivery of the programme is led by health visitors (if <5y old) and school nurses (if 5–19y) but involves the whole primary healthcare team as well as other care providers, e.g. children's centres/social care. Liaise with the health visitor/school nurse if:

- Immunizations are not up to date
- You have any worries regarding parenting abilities
- A child does not attend primary or secondary care appointments
- You have concerns about neglect or abuse (but also see ➔ p. 902)

Further information

Department of Health and Social Care (2009) Healthy Child Programme: Pregnancy and the First 5 Years of Life. 🖱 www.gov.uk/government/publications/healthy-child-programme-pregnancy-and-the-first-5-years-of-life

Department of Health and Social Care (2009) Healthy Child Programme: 5 to 19 years old. 🖱 https://www.gov.uk/government/publications/healthy-child-programme-5-to-19-years-old

Childhood screening Aims to discover physical, developmental, or behavioural problems as early as possible so that they can be managed to prevent secondary complications. It involves antenatal screening (➔ p. 782), neonatal bloodspot screening (➔ p. 830), newborn hearing screening (➔ p. 836), and the neonatal and 6–8wk checks (checking eyes, heart, and hips as well as developmental milestones—➔ p. 828).

Further developmental reviews are carried out by the health visitor, and the school health service checks height, weight, vision, and hearing. Any consultation can opportunistically be used to check immunization status, monitor development, and for health promotion. Expected developmental milestones are summarized in Table 23.3 (➔ p.832).

Diploma in Child Health Designed to give recognition of competence in the care of children to GP vocational trainees, staff grade doctors, and trainees in specialties allied to paediatrics. Administered by the Royal College of Paediatrics and Child Health (RCPCH). Further details are available at 🖱 www.rcpch.ac.uk

Health education for new parents

Reducing the risk of sudden infant death syndrome (SIDS)

- Stop smoking in pregnancy and do not let anyone smoke in the same house as your baby
- Breastfeed your baby
- Place your baby on his/her back to sleep
- Do not let your baby get too hot
- Do not suddenly stop using a dummy before your baby is 6mo old if your baby is used to having one
- Keep your baby's head uncovered—place your baby with his/her feet to the foot of the cot to prevent wriggling down under the covers
- It is safest for your baby to sleep in a cot in your bedroom for the first 6mo
- Sharing a bed with your baby is associated with SIDS, the risk is ↑ if your baby was premature or low birth-weight and if either parent:
  - Is a smoker—no matter where or when he/she smokes
  - Has been drinking alcohol
  - Takes medication or drugs that might make him/her drowsy
  - Feels very tired
- It is dangerous to sleep with your baby on a sofa, armchair, or settee
- If your baby is unwell, seek medical advice promptly

Protecting your baby from accidents and infections

- Keep small objects out of your baby's reach
- Stay with your baby when he/she is eating or drinking
- Make sure your baby's cot and mattress are in good condition and that the mattress fits the cot properly
- Install at least one smoke alarm
- Plan a way to escape a fire with your baby
- Never leave your baby alone in a bath or near water
- Immunize your baby
- Make sure your baby cannot reach hot drinks or the kettle or iron flex
- Only use toys suitable for your baby's age
- Never shake your baby—ask for help if their crying gets too much for you
- Use a properly fitted baby car seat that is the right size for your baby
- Do not use a baby walker
- Wash your hands before feeding your baby and make sure your baby's bottle and teats are properly sterilized

Benefits for parents and children

- **Maternity, paternity, and shared parental benefits** ➔ p. 765
- **Child Benefit** Available to anyone responsible for the upbringing of a child aged <16y (those who claim and are on higher income incur an income tax charge). Claim forms are in packs given to new mothers or can be downloaded from 🖱 www.gov.uk/child-benefit
- **Low-income benefits** ➔ p. 104
- **Free prescriptions and dentistry** All children <16y (18y if in full time education) and mothers <1y postpartum, and some families on low income are entitled to free prescriptions and dentistry—➔ p. 113

The neonatal and 6-week check

It is essential that a full neonatal check is carried out <72h after delivery. Most neonatal checks are carried out in maternity units before discharge, but checks may be provided by GPs as a National Enhanced Service if:

- The baby is discharged <24h after delivery
- The birth occurs at home or in a GP unit, or
- There is rapid discharge from an obstetric to a peripheral unit

All babies should also have a 6–8wk check. This is usually performed in the GP surgery and provided in GMS practices as an '*Additional Service*' (opting out results in a 0.7% ↓ in Global Sum).

Parental concerns Discuss any worries the parent(s) might have about the child. Review FH, pregnancy, and birth. At the neonatal check, arrange hepatitis B vaccination if the mother is hepatitis B +ve (➜ p. 797) or BCG vaccination if in a high-risk group (➜ p. 296).

Additional history for the neonatal check

- Has the baby passed urine? For boys, is the stream good? If no urine in the 1st 24h suspect renal abnormality and admit for further investigation. If poor stream, suspect posterior urethral valves, phimosis, or hypospadias
- Has the baby passed meconium? If no meconium in the first 24h, suspect meconium ileus and admit for further investigation

Physical examination Check the baby systematically—Table 23.1.

Moro reflex Elicit if concerned. Support head and shoulders about 15cm from the examination couch. Suddenly allow the baby's head to drop back slightly. The response—extension of the arms followed by adduction towards the chest should be brisk and symmetrical. This reflex disappears by 6mo.

Check vitamin K has been given

- Discuss any concerns with the parent(s)
- Deficiency of vitamin K can → *haemorrhagic disease of the newborn* with potentially serious effects, including death
- All parents should be offered IM vitamin K for their baby; if IM vitamin K is declined, they should be offered oral vitamin K. ❗ One dose of oral vitamin K does not confer full protection. Formula feeds contain vitamin K supplements but breastfed babies require further doses—ensure that they get them
- Babies at high risk of bleeding (premature, low birthweight, unwell babies, and those who have undergone instrumental deliveries)—should always have IM vitamin K

Health education At the neonatal check, discuss neonatal blood spot screening (➜ p. 830). In all cases, discuss (➜ p. 827):

- Feeding and nutrition
- Sleeping position
- Baby care
- Sibling management
- Crying and sleep problems
- Transport in a car

Features of common genetic disorders ➜ p. 840

6-week developmental milestones Table 23.3, ➜ p.832

Table 23.1 Check list for the neonatal and 6–8wk examination

General appearance		
Syndrome? Clusters of features, e.g. features of Down's/Turner's syndrome	Weight: small or large for gestation? Pallor, jaundice, or cyanosis. ❗ Slight peripheral cyanosis is normal	Skin: birthmarks; meconium staining; purpura; lanugo or evidence of post-maturity
Head and facial features		
Head circumference Caput succedaneum or cephalohaematoma Fontanelles—number (if 3, ?Down's syndrome), size, and tension	Accessory auricles Ptosis Sticky eye, subconjunctival haemorrhage, conjunctivitis? Cataract or red reflex?	Sternomastoid swelling Cleft lip Potter's facies Pierre Robin jaw (receding jaw with cleft palate)
Mouth		
Cleft palate? (➔ p. 910) Tongue tie? (➔ p. 843)	Profuse saliva (associated with oesophageal atresia)	Epstein's pearls
Arms and hands		
Proportion of arms/fingers Oedema	Palmar creases Fingers—number, webbing, deformity	Normal movements? Erb's or Klumpke's palsy (➔ p. 838)
Chest		
Distortion Breast enlargement	Respiratory rate[a] Recession	Air entry/added breath sounds
Cardiovascular examination		
Pulses (femoral/brachial)	Heart rate, rhythm, and sounds	Murmurs (➔ p. 858)
Abdomen		
Umbilical infection/discharge, or hernia	Anus: patency/position	Masses[b]
Genitalia		
♂: penis—size and shape; position of urethral orifice; testes (normal, undescended, or maldescended), hernia, or hydrocele ♀: clitoromegaly; vaginal bleeding; posterior vaginal skin tag (common)		
Back, legs, and feet		
Sacral pit/spina bifida (➔ p. 879) Scoliosis (➔ p. 453)	Hips (➔ p. 834) Proportion of feet/legs/body	Club foot (➔ p. 471) Toes—number, webbing, deformity
CNS		
Is the baby behaving normally?	Is the cry normal?	Are all 4 limbs moving equally and is the Moro reflex (if done) symmetrical?

[a] Respiratory rate <60 breaths/min is normal.

[b] In a normal infant, the liver is usually palpable as are the lower poles of the kidneys; the spleen and bladder are never palpable.

Neonatal bloodspot screening

Neonatal bloodspot screening, using a heel-prick blood sample, is carried out when babies are 5–8d old. Blood is placed on special filter paper and analysed to detect a range of conditions for which evidence shows that early treatment improves outcome. Test results are available by 6wk.

⚠ If screening is declined, it is important to flag in the child's notes that the child has not been screened in case the child becomes ill later on.

Sickle cell disease (➔ p. 645) UK incidence 1 in 2000 babies.

- Sickle cell disease ↑ risk of severe overwhelming infection and splenic sequestration crises. Early diagnosis allows prophylaxis with penicillin and vaccines, and parent training to enable early presentation for treatment when complications arise
- Screening detects abnormal haemoglobin. If found, a confirmatory test is performed on the original spot using a different technique. As well as babies with sickle cell disease, testing detects babies with sickle cell trait, other heterozygous states, and other haemoglobin abnormalities (e.g. haemoglobin E, thalassaemia). Even if these have no clinical consequences, current policy is to inform parents of the results. Ensure parents understand the meaning/significance of results
- If a child tests +ve, it is important that parents and siblings receive genetic counselling and are offered genetic testing for the condition

Cystic fibrosis (CF) (➔ p. 300) UK incidence 1 in 2500 babies.

- Screening detects immunoreactive trypsin (IRT) which is ↑ in children with CF. If IRT is ↑, the blood is then DNA tested for the most common gene alterations. Screening will also detect healthy carriers
- If a child tests +ve, offer parents and siblings genetic counselling and testing. If both parents are carriers of a CF gene, there is a 1 in 4 chance of any subsequent child being affected

❗ Not all gene mutations are tested for. Some babies with CF will be missed by newborn screening. Continue to watch for later presentations.

Congenital hypothyroidism UK incidence 1 in 3000 babies (♀ > ♂).

- Untreated, children with abnormally low levels of thyroid hormone fail to grow properly and have mild to severe learning disability
- The bloodspot is used to detect low levels of blood thyroxine
- Treatment with thyroxine replacement results in normal growth and development. Usually thyroxine replacement is needed lifelong

Inherited metabolic diseases 6 autosomal recessive, inherited metabolic conditions (Table 23.2). Defective genes result in enzyme deficiencies which lead to a build-up of amino- or organic acids resulting in developmental delay ± metabolic crises (poor feeding, nausea, vomiting, and drowsiness, → seizures, coma, and death if untreated).

Management Specialist care is essential. Treatment is usually with dietary restriction/supplements and can result in normal growth and development. Parents usually have clear instructions about what to do in the event of a metabolic crisis; prompt specialist care is always required—admit. Genetic counselling and screening of parents/siblings is important.

Table 23.2 Metabolic conditions included for blood spot screening

Condition	UK incidence	Features
Phenylketonuria (PKU)	1 in 10,000	● Children are unable to break down phenylalanine, an amino acid present in many foods. The baby appears normal at birth but develops severe developmental delay, learning difficulty, and seizures in infancy ● The bloodspot test detects high levels of blood phenylalanine
Medium-chain acyl-CoA dehydrogenase deficiency (MCADD)	1 in 10,000	● Inability to metabolize fats effectively. If fasting/infection, metabolic crises occur ● The bloodspot test detects high levels of C8 carnitine (a fatty acid of medium length)
Glutaric acidaemia type 1	1 in 100,000	● Infants are unable to break down glutamic acid formed from lysine and tryptophan ● Leads to poor growth, low muscle tone, dystonia, developmental delay, brain damage and metabolic crisis, usually aged <1y
Maple syrup urine disease	1 in 185,000	● Enzyme defects lead to build up of leucine, valine, and isoleucine ● Infants appear normal at birth but maple syrup odour and metabolic crises usually occur in <1wk. Milder variants may present later with poor growth and developmental delay
Homocystinuria	1 in 250,000	● Enzyme deficiency (several types) results in high levels of homocysteine in blood and urine ● Causes skeletal features (e.g. tall stature, high palate, pectus excavatum/carinatum), osteoporosis, hypopigmentation, optic lens dislocation, learning disability, seizures, stroke, psychiatric disorders
Isovaleric acidaemia	1 in 250,000	● Defect in leucine metabolism ● Acute form presents in days/weeks with distinctive odour, poor feeding, weight ↓, and metabolic crises. Mild form leads to periodic attacks of severe ketoacidosis

! Occasionally other metabolic diseases can also be detected, e.g. galactosaemia may lead to high levels of phenylalanine.

Further information

Public Health England (2017) Newborn blood spot screening programme: supporting publications. 🖱 www.gov.uk/government/collections/newborn-blood-spot-screening-programme-supporting-publications

Support for parents

Metabolic Support UK ☎ 0800 652 3181 🖱 https://www.metabolicsupportuk.org/

National Society for Phenylketonuria (NSPKU) ☎ 030 3040 1090 🖱 www.nspku.org

Summary of developmental milestones

Table 23.3 Summary of developmental milestones

Development	6-week check	8 months	18 months	3 years	4 years
Gross motor	Controls head when pulled to sitting position (0–3mo) Moro reflex (0–6mo)—should be absent >6mo Holds head in line/slightly higher than body with hips semi-extended during ventral suspension (0–10wk) Lifts head momentarily when lying prone (from birth)	Bears weight on legs (3–7mo) Can be pulled to sit (14wk–6mo) Sits with support (4–6mo) Sits without support (5–8mo) Crawls (6–9mo)	Gets to sitting position (6–11mo) Pulls to standing (6–10mo) Walks holding onto furniture (7–13mo) Walks alone (10–15mo) Walks backwards (12–22mo) Climbs stairs (14–22mo)	Climbs and descends stairs Runs (~15mo) Pedals tricycle (21mo–3y) Jumps in 1 place (21mo–3y) Kicks a ball (15–24mo) Stands on 1 foot for 1 second (22mo–3¼y)	Hops forward on 1 foot for 2m (3–5y) Stands on 1 foot for 5s (2¾–4½y) Walks heel-to-toe (3½–5¼y)—backwards (4–6y) Bounces and catches a ball (3¼–5½y)
Fine motor/vision	Stares (from birth) Follows horizontally to 90° (0–6wk)	Reaches out to grasp (palmar grasp) (3–6mo) Transfers and mouths (passes an object from 1 hand to the other and puts it in their mouth) (18wk–8mo) Fixes gaze on small objects (5–8mo) Follows fallen toys (4–8mo)	Points with index finger Casts (throws) (9–15mo) Delicate pincer grasp (10–18mo) Holds two bricks and bangs them together (7–13mo) Scribbles (12–24mo) Builds a tower of 3–4 bricks (16–24mo)	Picks up 'hundreds and thousands' Imitates a vertical line (18–33mo) Copies a circle (2¼–3½y) Threads beads Builds a tower of 8 bricks (21mo–3½y) Matches 2 colours	Copies a cross (3–4½y) and square (4–5½y) Draws a person with 3 parts (with all features—4½–6y) Recognizes colours (3–4¾y)

Hearing and speech	Responds to rattle or bell (from birth) Startle response (from birth)	Vocalizes (4–6mo) Polysyllabic babbling (6–10mo) Laughs (2–5mo) Responds to own name (4–8mo)	Turns to sound of name Jabbers continually Uses ‘mama’ and ‘dada’ (11–20mo—half by 15mo) Can say 3 words other than ‘mama’ and ‘dada’ (10–21mo) Points to eyes, nose, and mouth (14–23mo) Obeys simple instructions (15mo–2½y)	Uses plurals (30mo–3¼y) Uses prepositions (3–4½y) Joins words into sentences (50% by 23mo; 97% by 3y) Gives own name	Speaks grammatically (2½y–4¼y) Counts to 10
Social behaviour/ play	Smiles (0–10wk—mean 5wk) Turns to look at observer’s face (from birth)	Puts everything into mouth (4–8mo) Hand and foot regard (4–8mo) Plays peek-a-boo (5½–10mo)	Holds spoon and gets food to mouth (14mo–2½y) Explores environment (13–20mo) Takes off shoes and socks (13–20mo)	Plays alone Eats with spoon and fork Puts on clothes (2¼–3½y—with supervision) Washes and dries hands Separates from mother easily (2–4y) Dry in the day (2–4y)	Shares toys Brushes teeth Dresses without supervision (3¼–5½y) Comforts friends in distress (5y)
⚠ *Warning signs*	No red reflex No visual fixation or following Failure to respond to sound Asymmetrical neonatal reflexes Excessive head lag Failure to smile	Not sitting Fisting Hand preference (<1y) Squint Primitive reflexes persist— Moro response, stepping, asymmetrical tonic neck reflex	Unable to walk, weight bear, and/or stand without support Persistence of hand regard ± casting. No pincer grip Absence of babbling or cooing; inability to understand simple commands	Unable to speak in simple sentences Unable to understand speech Persistent toe-walking	Speech difficult to understand due to poor articulation or because of omission or substitution of consonants (confusion of ‘s’, ‘f’, and ‘th’ disappears by 6½y)

Screening for hip dysplasia

Developmental dysplasia of the hip (DDH) ranges in severity from dysplasia with dislocation or subluxation, through instability, to mild acetabular dysplasia with a stable hip joint. Incidence is estimated at 1–3% of newborns when all grades of severity are included, although fewer have dislocation (3 in 2000 live births); ♀:♂ ≈5:1. Associated with breech presentation. Often there is a FH. 20% of cases are bilateral.

Presentation

- High-risk babies (breech presentation at 36wk even if turns prior to delivery and/or FH) are routinely screened with USS in the first 6–8wk of life, even if newborn examination is normal
- Otherwise, usually detected by clinical examination as part of routine screening. Screening should take place <72h after birth and at the 6wk check. Screening tests should be taught *in vivo* by someone experienced in the technique
- Despite screening, some cases present late as toddlers with limp/ waddling gait; frequent falls; asymmetric thigh creases or limited hip abduction. Be alert for signs and take parental concerns seriously
- Some (particularly mild cases) go unnoticed until adulthood when they present with pain (from damage to the acetabular labrum) or premature osteoarthritis

Screening a child <3mo of age

- Screening tests should be performed in a warm room with the baby undressed and lying on a firm surface
- Flex hips and knees to 90° using one hand for each leg with thumbs on the inner side of the baby's knee and ring and little fingers behind the greater trochanters (Figure 23.1)
- Each hip is tested separately. The examiner's hand on the opposite side from the hip being tested is used to stabilize the pelvis. Hold the thumb over the symphysis pubis and fingers under the sacrum
- Only test once as repeated testing can damage the hips

Ortolani manoeuvre (Figure 23.1) Each hip is gently abducted while lifting the greater trochanter forward. As a dislocated hip is abducted, a clunk or jumping sensation is felt. It is difficult to tell the difference between a click of a normal hip and a clunk of an abnormal one—so refer any clicky or clunky hips for further investigation (usually USS or orthopaedic review). Hip abduction of <60° in 90° of flexion is also a sensitive sign.

Barlow manoeuvre This establishes whether the hips are dislocatable. Holding the legs as described previously, gently apply pressure along the line of the femur pushing it backwards out of the acetabulum. The judder of the femoral head slipping in and out of the acetabulum can be felt if the hip is dislocatable.

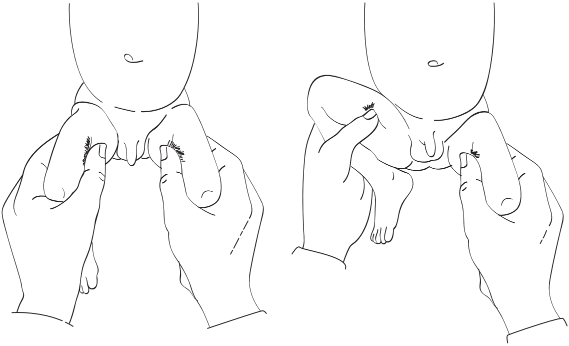


Figure 23.1 Screening for congenital dislocation of the hip (Ortolani test)

Examination of a child of >3mo of age

- >3mo of age, limited abduction is the most common finding in children with DDH. If the infant lies on his/her back with hips flexed at 90°, any hip which cannot abduct >75° should be viewed with suspicion
- Ortolani and Barlow tests are difficult to perform in older babies; X-ray is more useful than USS once a baby is >4.5mo old

Other signs

- Limb shortening on the affected side—compare knee levels with the child lying on his/her back and hips and knees flexed to 90°
- Asymmetry of buttock and posterior thigh skin folds when baby is held in ventral suspension
- Flattening of the buttock—in a prone position, the affected side may look flatter

Management Refer for an USS within 2wk if concerns at the neonatal examination. If concerns at 6–8wk check (or later) refer urgently to an orthopaedic surgeon specializing in paediatric problems.

Treatment depends on when the condition is diagnosed:

- Young babies Splinting in a pelvic harness to reduce and hold the hip—the hips are held in partial abduction using slings under each thigh attached to a body harness, e.g. von Rosen splint. Usually babies wear a splint for ~3mo
- Older babies, toddlers, and adults Surgery is required

Support for parents and children

Steps Support for patients with lower limb conditions and their families.
☎ 01925 750271 🖱 www.steps-charity.org.uk

Vision and hearing screening tests

Operational senses are essential for normal development. Conditions which interfere with the senses, even if correctable, may lead to permanent impairment if not detected and treated early.

Vision screening Carried out at the neonatal and 6wk check:

- Ask about parental worries about the child's vision
- Is there a FH of visual disorders (particularly retinoblastoma or congenital cataracts)?
- Inspect the external eye—are there any abnormalities?
- Check the red reflex in each eye (urgent referral if absent)
- At 6wk observe eye movement and fixing and following
- Children age 4–5y are offered vision screening at school, aiming to detect refractive errors and amblyopia

Refer children for further assessment at any age where there is concern about vision on assessment or parental concern.

⚠ *Warning signs for visual problems*

- The child does not fix on the mother's face while feeding by 6wk
- In a child >6wk old, the child's eye wanders about from one side of the eye socket to the other while the child is awake and happy
- A white spot is seen in the pupil at any age—could be cataract
- A child holds objects close to the face while trying to look at them
- A child >6mo old has a squint in one or both eyes

Tests for squint

- Sit the child on the parent's lap; stand in front of the child and shine bright light (e.g. pen torch) at arm's length from the child
- Fix the child's head in the midline and look for the reflection of the light on the child's corneas; the reflection should be symmetrical and near the centre of the pupil (usually slightly towards the nose)
- Turn the child's head to one side keeping the eyes fixed on the light—the reflection should stay symmetrical; repeat, turning to the other side
- If reflections are not symmetrical, perform a cover test

Cover test

- Sit the child comfortably on a parent's lap and shine a bright light or a place a small bright object at arm's length from the child
- Cover one eye with a card—watch for any movement of the uncovered eye to fix on the object. Then remove the card and watch the covered eye to see if it moves to fix on the object. Repeat with the other eye
- If either or both eyes move, a squint is present—refer

Hearing tests All newborn babies in the UK are offered hearing tests through the neonatal hearing screening programme (NHSP):

- **Automated oto-acoustic emission (AOAE) testing** Measures integrity of the inner ear and is offered to all neonates. An earpiece is placed in the ear and quiet clicking sounds are played. In a hearing ear, the cochlea produces sounds in response to the clicks that can be analysed. Screening takes a few minutes and can be done at the bedside when the baby is asleep but responses may be unclear especially if the baby is aged <24h

- **Automated auditory brainstem response (AABR) testing** Assesses the entire auditory pathway and is offered to any baby not passing the AOAE test or to any baby that has a stay of >48h in a special care baby unit (in addition to AOAE testing). Sensors are placed on the head/neck and quiet clicking sounds are played through headphones. Responses to sounds around the brain stem are analysed

⚠ *Warning signs for hearing problems*

- No startle response to loud noises at 6wk
- The child does not respond to his/her name by 8mo
- Absence of babbling or cooing by 1y
- Inability to understand simple commands by 18mo
- Inability to speak in short sentences by 2½y

When are further hearing tests needed? Ensure referral to audiology for further assessment:

- **If the newborn hearing test indicates a problem**
- **At any age** If there is parental/professional concern; after temporal bone fracture; after bacterial meningitis/septicaemia (test <4wk after discharge from hospital); severe unconjugated hyperbilirubinaemia
- **At 8mo of age** If the child moved to the UK after 3mo of age; congenital infection (e.g. CMV, toxoplasmosis, rubella); neurodegenerative/neurodevelopmental disorder; syndrome associated with hearing loss (e.g. Down's or Turner's syndrome); craniofacial abnormality (e.g. cleft palate)

Delayed speech development May be due to global or specific learning disability, deafness, or neurological problems. Parents often compare their children's development to others leading to unnecessary anxiety. The range of normal for speech development is wide:

- First words 11–20mo
- A 2y old may use anything from a few words to 2000 words
- Children start using prepositions at any time from 3–4½y

Management If a child's speech is delayed, check other developmental milestones; examine for any neurological deficit and check hearing. Consider autism (➔ p. 896) particularly if regression of speech. Refer to paediatric speech therapist in accordance with local guidelines.

Further information

Public Health England (2016) Newborn hearing screening: programme overview. 🖱 www.gov.uk/guidance/newborn-hearing-screening-programme-overview

Royal College of Ophthalmologists Ophthalmic services for children. 🖱 www.rcophth.ac.uk/wp-content/uploads/2014/12/2012_PROF_182_Ophthalmic-Services-for-Children.pdf

Parent and child information and support

LOOK Support for families of blind or visually impaired children. ☎ 01432 376314 🖱 www.look-uk.org

National Deaf Children's Society ☎ 0808 800 8880 🖱 www.ndcs.org.uk

Birth trauma

Head trauma

Caput succedaneum Swelling, bruising, and oedema of the presenting portion—usually scalp. Unsightly but resolves spontaneously.

Cephalohaematoma Uncommon. Haemorrhage beneath the periosteum. Unilateral and usually parietal. Presents as a lump—the size of an egg—on the baby's head. Treatment is not required, but anaemia or hyperbilirubinaemia may follow.

Depressed skull fracture Rare. Most result from forceps pressure; rarely caused by the head resting on a bony prominence *in utero*. May be associated with subdural bleeding, subarachnoid haemorrhage, or contusion/laceration of the brain itself. Seen and felt as a depression in the skull. X-ray confirms diagnosis; may need neurosurgical elevation.

Intracranial haemorrhage Rare. Suggested by lack of responsiveness, fits, respiratory distress ± shock. Admit as an emergency.

Nerve injuries

Cranial nerve trauma The facial nerve is injured most often, causing facial asymmetry especially during crying. Usually resolves spontaneously by 2–3mo of age.

Brachial plexus injury Follows stretching caused by shoulder dystocia, breech extraction, or hyperabduction of the neck in cephalic presentations. Often associated with other traumatic injuries, e.g. fractured clavicle or humerus, subluxations of the shoulder or cervical spine.

- **Partial injuries of the brachial plexus** Most recover but site and type of nerve root injury determine prognosis. If persists, refer to paediatric neurology for further investigation
  - Injuries of the upper brachial plexus (C5–6) affect muscles around the shoulder and elbow—*Erb's palsy*
  - Injuries of the lower plexus (C7–8 and T1) affect primarily muscles of the forearm and hand—*Klumpke's palsy*
- **Injuries of the entire brachial plexus** No movement of the arm + sensory loss. Refer immediately for neurological opinion. Prognosis for recovery is poor

Fractures

Mid-clavicular fracture Most common fracture during birth. Usually occurs due to shoulder dystocia. Most clavicular fractures are greenstick fractures and heal rapidly and uneventfully. A large callus forms at the fracture site in <1wk and remodelling is completed in <1mo. Can be associated with brachial plexus injury and/or pneumothorax.

Long bone fractures The humerus and femur may be fractured during difficult deliveries. Usually long bones heal rapidly without any residual deformity.

Cerebral palsy^{N} The term cerebral palsy identifies children with non-progressive spasticity, ataxia, or involuntary movements. Mixed patterns are common. It affects 0.2% of children.

Risk factors/causes

- Prematurity (~1%)
- Low birthweight (~1%)
- Congenital malformations
- Maternal infections
- Neonatal jaundice
- Early childhood meningitis/sepsis
- Perinatal asphyxia
- CNS trauma

Associated disorders

- Epilepsy (33%)
- Visual problems (50%)
- Learning disability (50%)
- Communication difficulties (50%)
- Hearing impairment (10%)
- Behavioural/psychiatric/attention problems

Spastic syndromes 70%. Upper motor neurone involvement.

- May cause hemiplegia, paraplegia, quadriplegia, or diplegia
- Affected limbs are underdeveloped and have ↑ tone, weakness, and a tendency toward contractures
- A scissor gait and toe-walking are characteristic
- With quadriplegia dysarthria is common

Athetoid/dyskinetic syndromes 20%. Basal ganglia involvement.

- Characterized by slow, writhing, involuntary movements affecting the extremities (athetoid) or proximal parts of the limbs/trunk (dystonic)
- Abrupt, jerky, distal movements (choreiform) may also occur
- Movements ↑ with emotional tension and stop during sleep
- Dysarthria is often severe

Ataxic syndromes 10%. Involvement of the cerebellum. Weakness, incoordination, and intention tremor produce unsteadiness, wide-based gait, and difficulty with rapid and fine movements.

Diagnosis Early signs include movement problems (fidgeting, ↓, or asymmetric movements), hypotonia or spasticity, delayed motor milestones, and feeding difficulties. If any of these features are present (especially if risk factors present), refer urgently for developmental assessment. Also refer any child not sitting by 8mo, walking by 18mo, with hand preference before 1y, or persistent toe-walking.

Management Aims to achieve maximum independence. Requires MDT approach and treatment of associated difficulties (e.g. feeding, sleep, toileting) and pain as well as motor problems. Child and parents need assistance in understanding disability, setting realistic goals, and managing their own feelings.

The chronically disabled child ➔ p. 900

Further information

NICE (2017) Cerebral palsy in under 25s: assessment and management.
🖱 www.nice.org.uk/guidance/ng62

Information and support for parents

SCOPE (cerebral palsy) ☎ 0808 800 3333 🖱 www.scope.org.uk

Genetic disorders

There are many thousands of genetic disorders that can affect children.

Modes of inheritance ➲ p. 176

Management of children with long-term disability ➲ p. 900

> ⚠ Some genetic syndromes are associated with cancer (e.g. Down's syndrome and leukaemia; neurofibromatosis and CNS tumours).

Structural chromosome syndromes Table 23.4

Fragile X syndrome Affects 1 in 4000 ♂ births and 1 in 6000 ♀ births. Carried on the X chromosome. Comprises:

- Low IQ (20–70)
- Large testes (♂)
- High forehead
- Large jaw
- Long ears
- Facial asymmetry
- Associated with autism, ADHD, anxiety, and OCD

50% of carrier ♀s have normal IQ; 50% have some learning disability. Consider fragile X syndrome in any child with developmental delay of unknown cause. Antenatal testing is possible for future pregnancies.

Tuberous sclerosis *Incidence*: 1 in 5000–10000. Autosomal dominant inheritance. Caused by mutations of *TSC1*/*TSC2* genes on chromosomes 9 and 16; >60% arise from new mutations. Characterized by hamartomatous skin, nervous system and internal organ lesions. Usual presentation is with adenoma sebaceum (angiofibromas of the skin—seen as red-brown papules on the face—appear aged 5–10y), epilepsy, and developmental delay.

Other features include: coarsened skin over the sacrum (shagreen patch); nail fold fibromas; hypopigmented oval skin patches (ash leaf spots); cardiac, renal, lung, and eye abnormalities. Treatment is supportive.

Tay–Sachs disease Autosomal recessive mutation of the *HEXA* gene on chromosome 15. *Incidence*: 1 in 320,000; 1 in 25 of Ashkenazi Jewish populations carry the gene. Refer for antenatal screening (➲ p. 785) if from high-risk population. Otherwise treatment is supportive. 3 forms:

- Infantile Most common. Symptoms appear at ~6mo with relentless deterioration in neurological function until death at <4y
- Juvenile Symptoms appear aged 2–10y; death occurs <15y
- Adult or late onset Rare. Neurodegenerative symptoms develop in adolescents/adults. Results in long-term disability

Glycogen storage diseases *Incidence*: ~1 in 25,000. Lack of enzyme(s) involved in glycogen synthesis/breakdown → deposition of abnormal amounts/types of glycogen in tissues. Autosomal recessive except for type IX (in which most cases are X-linked). Symptoms and age of onset vary considerably according to predominant organs involved:

- Liver (I, III, IV and VI) → hepatomegaly, hypoglycaemia, metabolic acidosis
- Muscle (types II, V, VII) → weakness, lethargy, poor feeding, heart failure

Treatment involves dietary modification (small carbohydrate meals ± high protein); allopurinol (to prevent renal urate stones and/or gout) ± limiting anaerobic exercise.

Screening for inherited metabolic disorders ➲ p. 830

Table 23.4 Common structural chromosome syndromes

Genetic problem	Features
Down's syndrome Trisomy 21 (92%) Translocation (6%) Mosaicism (2%) *1 in 1000 live births*	*Facial abnormalities*: flat occiput, oval face (mongoloid facies), low-set eyes with prominent epicanthic folds *Other abnormalities*: single plantar crease, hypotonia, congenital heart disease Developmental delay Life expectancy is ↓ but >50% live to >60y
Edward's syndrome Trisomy 188 *1 in 6000 live births* ♀:♂ ≈2:1	*Facial abnormalities*: low-set, malformed ears, receding chin, protruding eyes, cleft lip/palate *Other abnormalities*: short sternum makes the nipples appear widely separated; fingers cannot be extended and the index finger overlaps the 3rd digit; umbilical/inguinal hernias; rocker-bottom feet; rigid body with flexion of limbs Developmental delay Life expectancy is ~10mo
Patau's syndrome Trisomy 13 *1 in 10,000 live births*	*Facial abnormalities*: small head and eyes; cleft lip/palate *Other abnormalities*: skeletal abnormalities, e.g. flexion contractures of hands ± polydactyly with narrow fingernails; brain malformation; heart malformation; polycystic kidneys 50% die in <1mo. Usually fatal in the first year
Cri-du-chat syndrome Deletion of short arm of chromosome 5 *1 in 50,000 births*	*Facial abnormalities*: microcephaly; marked epicanthic folds; moon-shaped face; alert expression; abnormal cry (cat-like) Developmental delay Usually fatal in the first year
Turner's syndrome XO—deletion of one X chromosome or mosaicism (XO. XX) *1 in 2000 ♀ births*	Female appearance *Facial abnormalities*: ptosis, nystagmus, webbed neck *Other abnormalities*: short stature (<130cm); hyperconvex nails; wide carrying angle (cubitus valgus); inverted nipples; broad chest; coarctation of the aorta; left heart defects; lymphoedema of the legs; ovaries rudimentary or absent Lifespan is normal
Klinefelter's syndrome XXY or XXYY polysomy *1 in 1000 ♂ births*	Male appearance. Often undetected until infertility in adult life. *Other features*: psychopathy; ↓ libido; ↓ facial hair; gynaecomastia (↑ risk breast cancer); small, firm testes *Associations*: hypothyroidism; DM; asthma

Information and support for families

Association for Glycogen Storage Disease (UK) 🖱 www.agsd.org.uk
Cure and Action for Tay–Sachs 🖱 www.cats-foundation.org
Down's Syndrome Association ☎ 0333 1212 300 🖱 www.downs-syndrome.org.uk
Fragile X Society ☎ 01371 875100 🖱 www.fragilex.org.uk
Genetic Alliance UK, Rare Disease UK and Swan UK ☎ 020 7831 0883 🖱 www.geneticalliance.org.uk
Soft UK 🖱 www.soft.org.uk
Tuberous Sclerosis Association 🖱 www.tuberous-sclerosis.org
Turner's Syndrome Support Society ☎ 0300 111 7520 🖱 www.tss.org.uk
Unique ☎ 01883 723356 🖱 www.rarechromo.org

Common problems of small babies

Minor problems of neonates and small babies Table 23.5

Crying ➔ p. 888

Weight loss[N] Some weight ↓ (<10%) is common in neonates; most regain birthweight by 3wk. Arrange assessment/support for feeding if weight ↓ is >10% of initial weight, or is not regained by 3wk. Refer to paediatrics if marked weight ↓, unwell, any abnormality on examination, or no improvement with feeding support. Monitor if not referred.

Posseting (gastro-oesophageal reflux)[N] Common (>40% of infants); frequent (>6×/d in 5%); usually starts aged <8wk; resolves in <1y in 90%. Reassure. If the baby is thriving and not distressed, no investigation/treatment is needed. ❗ Warn parents to seek urgent medical review if 'red flag' symptoms: projectile, bile, or bloodstained vomiting.

Gastro-oesophageal reflux disease[N] Posseting/gastro-oesophageal reflux causing distress or complications (e.g. faltering growth, aspiration pneumonia, frequent otitis media). More common if premature or neurodisability (e.g. cerebral palsy). ❗ Cow's milk protein allergy (➔ p. 867) can present in a similar way. *If associated with distress*:

- **Breastfed babies** Arrange feeding assessment; consider alginate therapy
- **Bottle-fed babies** Review feed volumes, ↓ if excessive; try smaller more frequent feeds. If unsuccessful, offer a 2–3wk trial of thickened feeds; stop thickened feeds if no improvement and offer alginate therapy

Refer to paediatrics if

- Continued distress or faltering growth despite simple measures
- Dysphagia, haematemesis/melaena, and/or unexplained anaemia
- Failure to improve by age 1y

Colic Common in newborns ≤3mo. Repeat bouts of intense, unstoppable crying, often in early evening. Body is tense/rigid, face red, and knees drawn up. Attributed to abdominal pain but no proof. Ask about general health, feeding, onset/duration of crying, parental response, relieving factors, and FH of allergy. Normal examination. Reassure that colic is common, usually no serious underlying cause, and the baby is not rejecting the parents. Holding the baby, gentle motion, white noise, or a warm bath may help. Consider a trial of hypoallergenic formula (➔ p. 848); colic drops do not help.

Refer to paediatrics if

- Diagnosis is in doubt
- Severe symptoms
- Other symptoms or signs (e.g. faltering growth, severe eczema), or
- Fails to resolve by 12wk of age

Further information

NICE (2006, updated 2015) Postnatal care up to 8 weeks after birth. 🖱 www.nice.org.uk/guidance/cg37

NICE (2015) Gastro-oesophageal reflux disease in children and young people: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng1

NICE (2017) Faltering growth: recognition and management of faltering growth in children. 🖱 www.nice.org.uk/guidance/ng75

Table 23.5 Minor problems of neonates and small babies

Condition	Features	Management
Milia	Tiny, pearly white papules on the nose/face	Disappear spontaneously—reassure
Epstein's pearls	Small yellow/white cysts on the palate ± gums.	Reassure—resolve spontaneously
Erythema toxicum (neonatal urticaria)	Red blotches with a central, white vesicle. Each spot lasts ~24h. Spots are sterile. Baby is well	If infection suspected, take a swab. Otherwise reassure; resolves spontaneously
Tongue tie (ankyloglossia)	4–11% of newborns ♂ > ♀ Frenulum connecting the tongue to the floor of the mouth is short, restricting tongue movement	If affecting feeding, refer for frenulotomy (the frenulum is cut without anaesthetic)
Harlequin colour change	One side of the body flushes red; the other half stays pale	Harmless vasomotor effect—reassure
Single palmar crease	Common abnormality associated with genetic syndromes, e.g. Down's	Of no consequence unless other abnormalities
Milaria (heat rash)	Itchy red rash which fades as soon as the baby is cooled	**Reassure.** Keep the baby cool if the rash appears
Peeling skin	Common among babies born after their due date	Apply moisturizer to prevent skin cracking
Petechial or subconjunctival haemorrhage	May all occur during delivery	Resolve spontaneously—reassure. Ensure the baby has had vitamin K supplements
Swollen breasts	Due to maternal hormones Occurs in both sexes Occasionally lactate 'witch's milk'	Breast swelling usually subsides spontaneously. May become infected and require antibiotics
Sneezing	Neonates clear amniotic fluid from their noses by sneezing	Reassure
Umbilicus	After birth, the umbilicus dries, becomes black, and separates at about 1wk of age	Umbilical stump can become infected (offensive odour, pus, periumbilical flare, malaise)—treat with antibiotics If granuloma forms at the site of separation, exclude a patent urachus (refer if present) and treat with silver nitrate cautery
Red-stained nappy	Common in the first few days of life. Usually due to urinary urates but may be due to blood from the cord or vagina (oestrogen withdrawal bleed)	Reassure
Sticky eye	Common. Usually due to a blocked tear duct. Swab to exclude ophthalmia neonatorum	Ophthalmia neonatorum—➔ p. 716. Blocked tear duct (➔ p. 943)—bathe with boiled water when changing nappies. Avoid antibiotics unless overt infection

Problems of prematurity

Any baby born at <37wk gestation is premature. Worldwide, 11% of babies are born prematurely (7.4% in the UK). Prematurity affects all systems of the body and in general the problems are worse the more premature the baby:

- **32–36wk gestation**—premature. Generally do well—many needing only tube feeding and warmth
- **28–32wk gestation** (1–2% of births)—very preterm. In the UK, >90% babies born at 28wk gestation survive
- **<28wk gestation** (0.4% of births)—extremely preterm. Some babies as premature as 23–24wk gestation survive, but high incidence of disability

Nutrition Preterm babies <34wk suck and swallow poorly so commonly need nasogastric tube feeding. They are at particular risk of hypoglycaemia and need frequent feeds. Breast milk—either the mother's or donated—is preferred, sometimes with calorie and mineral supplements. If this is not available, special low-birthweight formula is used. Vitamin and iron supplements are routine until >6mo of age. Gastro-oesophageal reflux is common. For extremely preterm babies, feeding problems can persist into childhood.

Thermoregulation Poor in preterm infants, as they have a high surface area-to-body weight ratio and little subcutaneous fat. A controlled temperature/adequate insulation with clothes and blankets, is important.

Respiration

- **Preterm infants >34wk gestation** May have transient tachypnoea at birth due to inability to express fluid from the lungs, and may need O_2 therapy
- **Preterm infants <34wk gestation** Insufficient surfactant may be produced causing *respiratory distress syndrome* and requiring surfactant replacement and continuous positive airway pressure (CPAP) or mechanical ventilation. Incidence and severity is ↓ by antenatal corticosteroids
- **Extremely premature babies** May develop chronic lung disease—defined as being ventilator or O_2 dependent at 36wk post conception—due to *bronchopulmonary dysplasia*. These babies may be sent home on O_2 via nasal cannulae. They are at higher risk from respiratory infections particularly RSV. Episodes of bradycardia and apnoea are common. Have a low threshold for readmission

! All babies born prematurely are at ↑ risk of wheezing, asthma, chest infections, and sleep apnoea—the more premature, the greater the risk.

Prevention of RSV infection

- Take precautions to prevent exposure, e.g. avoiding busy waiting rooms
- Palivizumab is a monoclonal antibody indicated for the prevention of RSV infection in infants at high risk of infection. Prescribe *only* under specialist supervision. Give the first dose before the start of the RSV season and then give monthly throughout the RSV season

Jaundice The immature liver is less able to process bilirubin so premature babies are at ↑ risk of neonatal jaundice and are more likely to develop kernicterus—have a low threshold to refer for phototherapy.

Infection The immune system is poorly developed, so there is greater risk of infection. Furthermore premature babies may exhibit few signs of infection and serious illness, so have a low threshold to refer to paediatrics for a septic screen and antibiotics.

Anaemia Low iron stores, ↓ red cell survival, low levels of erythropoietin, and repeated venepuncture all lead to anaemia in premature babies. Some very premature babies need repeated transfusion. Iron supplements are routinely given to most premature babies and should be continued until >6mo old.

Neurodevelopment[N] Preterm birth is associated with:

- ↑ risk of cerebral palsy (➔ p. 838) and other motor function problems
- Learning disability/low educational achievement
- Speech and language difficulties
- ADHD
- Autism spectrum disorder (➔ p. 896)
- Emotional and behavioural problems

For most of these, risk ↑ with ↓ gestational age, with significant periventricular haemorrhages and cystic leucomalacia, and sepsis being independent risk factors. Some of these problems may not be apparent until well beyond the neonatal period and enhanced developmental screening (arranged by the neonatal units) is recommended.

Vision *Retinopathy of prematurity* (abnormal vascularization) may occur in very premature babies and may result in visual impairment—even blindness. All babies born at <32wk gestation or <1501g birthweight should have ophthalmological examination prior to discharge from hospital but may need follow-up and/or laser treatment once home.

Hearing Babies requiring neonatal intensive or special care have ↑ risk of hearing problems (10–20× ↑ prevalence of significant bilateral hearing loss). They should have neonatal screening prior to discharge from hospital and appropriate follow-up.

Bonding Separation of mother and premature baby is often necessary. Poor bonding is common—and the problem is added to by fear of losing the baby. In many special care units, periods of 'kangaroo mother care' are used to improve bonding—the baby is nursed skin-to-skin attached to the mother/father's chest. Parents may be (quite understandably) anxious when their babies first come home after a long period in special care and need more support and reassurance than other new parents.

Sudden infant death syndrome (SIDS) Premature babies have ↑ risk of SIDS. *Prevention*: ➔ p. 827; *Management*: ➔ p. 904

Further information

NICE (2017) Developmental follow-up of children and young people born preterm. 🖱 www.nice.org.uk/guidance/ng72

Information and support for parents of premature babies

Bliss Support Line. ☎ 0808 801 0322 🖱 www.bliss.org.uk

Neonatal jaundice

In the first few days of life, most babies have ↑ serum bilirubin levels as the liver takes over the excretion of bilirubin from the placenta. Mild jaundice from age 2–6d is physiological and harmless. However, very high levels of unconjugated bilirubin are toxic, crossing the blood–brain barrier and causing encephalopathy (*kernicterus*).

High-risk babies Require an extra visual check for jaundice in the first 48h of life. These include babies:

- Born at <38wk gestation
- Exclusively breastfed
- With siblings who required phototherapy

⚠ **Red flag features** If a baby is unwell and is jaundiced, refer for same-day paediatric review regardless of age and without checking bilirubin level. Refer urgently to paediatrics if a baby has jaundice and pale, chalky stools, and/or dark yellow urine that stains the nappy (baby urine should be colourless); or if conjugated bilirubin is >25micromol/L.

Jaundice <24h after birth Refer any baby with visible jaundice in the first 24h immediately back to paediatrics to exclude pathological causes (usually haemolysis or infection) and for monitoring/possible treatment.

Significant jaundice >24h and <2wk after birth May be difficult to assess (particularly in a dark-skinned baby)—examine the baby naked in natural light, check the sclera of the eyes, gums, and press lightly on the skin to examine for jaundice on blanched skin.

Management of suspected jaundice Measure and record bilirubin level within 6h with a transcutaneous bilirubinometer (usually via midwife or paediatrics). Check serum bilirubin if bilirubinometer reading is >250micromol/L and/or above referral threshold (Table 23.6); baby is <35wk gestation; or a bilirubinometer reading cannot be obtained.

Table 23.6 Bilirubin referral thresholds for babies born >38wk gestation[N]

Age (h)	Bilirubin level (micromol/L)	
	Repeat test in 18h or 24h if:	Refer for phototherapy if:
24	>150	>200
30	>162	>212
36	>175	>225
42	>187	>237
48	>200	>250
54	>212	>262
60	>225	>275
66	>237	>287
72	>250	>300
78	>262	>312
84	>275	>325
90	>287	>337
96+	>300	>350

! Treatment thresholds differ according to gestational age at birth. Graphs for all gestational ages are available to download from www.nice.org.uk/guidance/cg98/resources

Management of raised bilirubin levels[N]

- If bilirubin level is above treatment threshold (Table 23.6) Refer to paediatrics for further investigation and phototherapy
- If bilirubin is <50micromol/L below treatment threshold (Table 23.6) Repeat bilirubin measurement in <18h if high risk or otherwise in <24h
- If bilirubin is ≥50micromol/L below treatment threshold And clinically well, do not routinely repeat bilirubin measurement

Jaundice persisting >2wk (>3wk in preterm babies) Although 10% of breastfed babies are still jaundiced at 1mo of age, it is important to refer to paediatrics to exclude pathological causes, including:

- Haemolysis
- Infection—particularly UTI (➲ p. 856)
- Neonatal hepatitis or biliary atresia
- Hypothyroidism

Neonatal hepatitis Presents with persistent neonatal jaundice. Always requires specialist investigation and management. *Possible causes*:

- Congenital infection, e.g. HBV
- Galactosaemia
- Cystic fibrosis
- Glycogen storage diseases
- α1-antitrypsin deficiency

Galactosaemia Autosomal recessive, inborn errors of metabolism characterized by ↑ plasma galactose. Clinical manifestations depend on enzyme defect(s):

- Galactokinase deficiency *Incidence*: 1 in 40,000. Presents in childhood with cataracts. Treatment involves a galactose-free diet
- Classic galactosaemia *Incidence*: 1 in 44,000. The child appears normal at birth but becomes anorexic and jaundiced within a few days or weeks of consuming breast milk or lactose-containing formula. Vomiting, poor growth, hepatomegaly, and septicaemia are common and can be rapidly fatal. Treatment involves eliminating all sources of galactose in the diet. Long-term complications—poor growth, learning difficulty, infertility, speech and neurological abnormalities—are common

Biliary atresia *Incidence*: 1 in 15,000. End-stage of a sclerosing process in an initially patent biliary tree. Cause is unclear. Presents with jaundice in neonates. Prognosis has improved with laparotomy and porto-enterostomy which relieves the problem in ~50–70% of babies. If unsuccessful, liver transplant is needed. ❗ Early diagnosis is particularly important so that surgery can be carried out <2mo after birth before the liver is irreversibly damaged.

Persistent physiological ('breast milk') jaundice Diagnosed when other causes of prolonged jaundice have been excluded. Reassure. Offer breastfeeding support if needed. Monitor until subsides.

Further information

NICE (2010, updated 2016) Jaundice in newborn babies under 28 days. 🖱 www.nice.org.uk/guidance/cg98

Support for parents

Children's Liver Disease Foundation ☎ 0121 212 3839 🖱 www.childliverdisease.org

Feeding babies

Breastfeeding Preferred way to feed infants from birth until fully weaned or longer. The UK has lowest breastfeeding rate in the world; 80% of mothers start breastfeeding but only 44% are feeding at 6–8wk. Problems with painful breasts/nipples, concern regarding amount of milk the baby is getting, and lack of support are common reasons for stopping. Breastfeeding is something some find natural and others find difficult. Teaching mother and baby to breastfeed takes time and patience. Be supportive; ask a midwife, HV, or local breastfeeding advisor to help if needed.

Advantages of breastfeeding

- Encourages a strong bond between mother and baby
- More convenient than bottle-feeding—the milk is ready warmed and there is no need for sterilized bottles
- Cheaper than bottle-feeding
- Protects the baby from infection
- ↓ risk of sudden infant death
- ↓ postpartum bleeding
- ↓ childhood obesity
- Possible ↓ in childhood atopy
- Possible ↓ risk of DM for baby and mother in later life
- Helps the mother ↓ weight after pregnancy, and space pregnancies
- Protects the mother against breast and possibly ovarian cancer

Common problems with breastfeeding Table 23.7

Bottle-feeding

- **Cow's milk formula feeds** Cow's milk altered to simulate the composition of human milk, with added iron and vitamins. Advise parents to choose a formula suitable for their baby and make it up exactly as the manufacturer directs. Advise parents to wash feeding bottles/teats well and sterilize them until >12mo of age. Use a cup rather than bottle >6mo
- **Follow-on formula** Contains more iron and casein. Not essential unless a child is not taking solids and is >6mo old. Baby milk suitable from birth can be used until a switch is made to normal cow's milk
- **Unmodified cow's milk** Not recommended until the baby is >1y old as less digestible and contains little iron
- **Hydrolysed protein and amino acid infant formula** (e.g. Nutramigen®, Neocate®, Aptamil® Pepti) are available on ACBS prescription if proven cow's milk protein allergy, lactose intolerance, or galactosaemia
- **Soya protein-based formula** Not recommended for babies <6mo of age because of concerns regarding phyto-oestrogen content. Prescribe only on consultant advice

Healthy Start Provides infant formula milk, fruit, vegetables, and free vitamins for mothers of babies <1y and for children aged 6mo–4y from low-income households—➔ p. 765.

Further information

UK Drugs in Lactation Advisory Service ☎ 0116 258 6491 or 0121 424 7298
🖱 www.ukmi.nhs.uk/activities/specialistServices/default.asp?pageRef=2
UNICEF Baby Friendly Initiative 🖱 www.unicef.org.uk/babyfriendly

Table 23.7 Common problems with breastfeeding

Problem	Possible solutions
Painful breasts and/or nipples	Ensure correct positioning; treat mastitis or thrush if present
Feeding difficult despite correct positioning	Consider tongue tie (➔ p. 843). If affecting feeding, refer for frenulotomy
It is difficult to know how much milk the baby is taking at each feed	Encourage demand feeding and tell mothers to exhaust milk supply in one breast before starting the other Plot weight. If there are concerns about weight ↑, consider other causes of faltering growth—➔ p. 850
Breast milk does not contain all the nutrients the baby needs	Breast milk has low levels of vitamin K, D, and iron Ensure babies who have had oral vitamin K at birth receive additional vitamin K supplements Encourage weaning at 6mo Vitamin D supplements (8.5–10 micrograms/d) are recommended for breastfed babies Iron drops can be given to babies with low iron reserves (e.g. low birthweight, maternal anaemia)
Only the mother can feed the baby	Mothers who anticipate they will be absent from the baby for a period of time can express milk for someone else to feed to the baby in a bottle while they are gone. Advise mothers not to attempt this before breastfeeding is well established as the baby might find the two techniques confusing Two methods are commonly used: • Using a commercially available breast pump, or • By hand into a sterile bowl Breast milk can be frozen (special bags are available) and defrosted when required. Bottles should be sterilized and the milk warmed in the same way as for bottle-feeding
Disease can be transferred in breast milk	In general, breast milk protects the baby from disease A few diseases can be transferred in breast milk, e.g. HIV; in those cases, bottle-feeding is recommended where uncontaminated water is available
Drugs taken by the mother may have adverse effects on the baby	Mothers should take medical advice before taking any drugs (including herbal remedies). For most conditions drugs safe for use while breastfeeding are available Rarely breastfeeding is contraindicated, e.g. chemotherapy

Sources of support for breastfeeding mothers

Association of Breastfeeding Mothers ☎ 0300 330 5453 🖱 www.abm.me.uk
Baby Café ☎ 0300 330 0700 🖱 www.thebabycafe.org.uk
Breastfeeding Network ☎ 0300 100 0212 🖱 www.breastfeedingnetwork.org.uk
La Leche League ☎ 0345 120 2918 🖱 www.laleche.org.uk
National Childbirth Trust (NCT) ☎ 0300 3300 700 🖱 www.nct.org.uk

Weaning and feeding problems

Breast and bottle feeding ➜ p. 848

Weaning Current guidelines recommend solids should be introduced at ~6mo, and while individual needs of infants and choices of parents should be considered and supported, never <17wk. Earlier introduction of solids is linked with ↑ rates of infection and ↑ incidence of allergy and intolerance to certain foods, e.g. gluten or eggs.

Advice for parents on weaning

- Continue with breast or formula milk—milk is the main source of food until the baby is a year old
- Babies are ready to be weaned when they can sit up; mouth objects; are interested in food and chewing; and can reach and grab accurately
- If starting solids when the baby is <6mo old, sterilize feeding bowls/cutlery before use, and avoid egg, gluten, and fish until the baby is >6mo of age
- Start with a few teaspoons of pureed or mashed fruit, vegetable, or cereal, or soft finger foods without added salt/sugar. Babies take time to learn how to feed from a spoon and it may be messy. Wait for the baby to open his/her mouth, and let babies touch the food
- Gradually offer a wide variety of different foods and textures. Do not give raw eggs or honey to babies <1y. Encourage 'finger foods' as soon as the baby can feed him/herself—try pieces of soft fruit, vegetable, or toast
- It is usual for stool to change consistency on weaning

Feeding problems Parents commonly complain that their child is not eating enough or is eating the wrong foods. Look for underlying problems if the child is not growing or developing as normal.

Normal growth/development Reassure the parents that it is normal for the amount of food eaten to vary from day to day. Consider referral to the health visitor for advice/support. Advise parents to:

- Sit down for family meals wherever possible, and chat about other things
- Restrict snacks/sugary drinks between meals, avoid using as a reward
- Give small portions, be patient, and praise children for eating
- Show little emotion if a child rejects a food, remove it without comment and try it again another time

Faltering growth (failure to thrive)^{N} Refers to slower childhood weight ↑ than expected for age and sex. Use UK-WHO growth charts, Thresholds for concern:

- A fall across 1 or more centiles if birthweight is <9th centile
- A fall across 2 or more centiles if birthweight was 9th–91st centile
- A fall across 3 or more centiles if birthweight was >91st centile

If weight is a concern, measure length. If aged >2y, measure height and calculate body mass index (BMI). A BMI <0.4th centile always requires further assessment/intervention; a BMI <2nd centile may need assessment but may also simply be due to a small build.

Non-organic causes Not always possible to identify, may be complex and multifactorial, interaction difficulties may be present but not be the main cause. Neglect is an uncommon cause of poor growth.

Organic causes

- Chronic infection
- Heart disease
- Metabolic disease, e.g. DM
- GI disease, e.g. coeliac
- Respiratory disease, e.g. chronic lung disease, cystic fibrosis
- Physical feeding problems, e.g. cleft palate, pyloric stenosis

Assessment Take a feeding history—quantities, times of the day, how feeds are made up. Ask about feeding problems (e.g. regurgitation of food, vomiting) and about other physical problems (e.g. breathlessness, diarrhoea). Examine the child carefully from top to toe looking for any physical abnormalities or signs of developmental delay. Consider an observation of feeds/mealtimes (e.g. by health visitor) and/or investigation for UTI/coeliac disease.

Management Try to use the same scales on each occasion. Treat any reversible causes. Provide feeding support and advice. Refer to or discuss with paediatrician if:

- Any abnormality requiring specialist paediatric care is found—urgency depends on the nature of the abnormality and degree of faltering growth
- Rapid weight ↓ or severely undernourished (urgent referral)
- Failure to respond to primary care treatment/intervention
- Unexplained short stature (➔ p. 870) or height gain (➔ p. 871)
- Safeguarding concerns (± refer to social services)

❗ Parents may feel blamed; involve fully in management/ongoing monitoring.

Growth disorders ➔ p. 870

Pyloric stenosis Infantile hypertrophic pyloric stenosis usually develops in the first 3–6wk of life (rare >12wk). Failure of the pyloric sphincter to relax results in hypertrophy of the adjacent pyloric muscle. Typically affects firstborn, male infants. Pyloric stenosis runs in families and is associated with Turner's syndrome, PKU, and oesophageal atresia.

Presentation

- Projectile vomiting of milk—no bile. The child is still hungry after vomiting and immediately feeds again. Rarely there is haematemesis
- Faltering growth
- Dehydration and constipation ('rabbit pellet stools')
- Pyloric mass (feels like an olive) is palpable in the right upper abdomen (95%)—especially if the child has just vomited
- Visible peristalsis in the epigastrium after a test feed

Differential diagnosis

- Posseting/reflux
- Overfeeding
- Gastroenteritis
- Milk allergy
- Infection—especially UTI
- ↑ intracranial pressure
- Uraemia
- Adrenal insufficiency
- Other causes of intestinal obstruction

Management Admit to paediatrics/paediatric surgery. Treatment is with a Ramstedt's pyloroplasty. There are usually no long-term effects.

Information and support for parents

Child Feeding Guide 🖱 www.childfeedingguide.co.uk
Parentline ☎ 0808 800 2222 🖱 www.familylives.org.uk
NHS England Your pregnancy and baby guide. 🖱 www.nhs.uk/conditions/pregnancy-and-baby

Fever and acute illness in the under 5s

Assessing sick children can be difficult. Infants <6mo old can be particularly difficult to assess and may deteriorate rapidly over a short period of time. Take parents' concerns seriously. Physical signs are often absent or deceptive. One approach is to exclude 'alarm' symptoms and signs that might point to serious illness. In general, the younger the baby, the lower your threshold should be for seeking a paediatrician's opinion.

Fever in older children Aged 5–12y ➔ p. 1058; aged ≥12y ➔ p. 1057.

Remote assessment[N] Ask about the features shown in Figure 23.2.

- If symptoms/signs suggesting life-threatening disease (e.g. compromised airway, breathing, or circulation, or ↓ level of consciousness)—arrange for immediate hospital transfer as a 'blue light' emergency
- Children with any Red features are at high risk of serious illness. If there are no features of an immediately life-threatening illness, arrange for review by a doctor in a face-to-face setting in <2h
- Amber features indicate intermediate risk of serious illness. Arrange for assessment by a doctor in a face-to-face setting the same day
- Children with all Green features and no amber or red features are at low risk of serious illness and often can be managed with advice

Face-to-face assessment[N] Take a history and perform a full physical examination, recording temperature, respiratory rate, heart rate, capillary return, and peripheral oxygen saturation (where possible). Think '*Could this be sepsis?*' Assess for features of serious disease (Figure 23.2). Remember to check under clothing and nappies for rashes. Localizing signs may be absent (e.g. tonsillitis may cause vomiting in small children).

- If any Red symptoms or signs are present, refer to a paediatrician for immediate or same-day review, depending on clinical state of the child
- If any Amber symptoms or signs are present, and a cause is found, treat the cause. If no diagnosis is made, decide on further action based on knowledge of the family and the clinical state of the child. 3 options:
  - Advise the child's parents to call to request medical review if there is any deterioration (give advice about symptoms/signs they should watch for) or if the child fails to improve within a defined period of time
  - Arrange to review the child again within a few hours, or
  - Refer for paediatric review
- If all Green features and no amber or red features are present, give advice about management at home and symptoms/signs that should prompt carers to seek further advice

❗ Do not prescribe antibiotics for fever of unknown cause or distinguish between serious/non-serious illness based on response to antipyretics.

Measuring temperature For infants <4wk old, use an electronic thermometer in the axilla. For children aged >4wk use an electronic or chemical dot thermometer in the axilla or infrared ear thermometer.

Common causes of pyrexia Childhood infections are the most common cause of fever among children in general practice (➔ p. 854). Consider UTI (➔ p. 856) if no localizing symptoms/signs. Think of TB and endocarditis—especially in high-risk patients. Do not forget tropical diseases, e.g. malaria in children returning from abroad.

Other causes of pyrexia (may present as prolonged fever) Include: malignancy (e.g. lymphoma, leukaemia), immunological causes (e.g. Still's or Kawasaki disease); drugs (e.g. antibiotics); and liver or renal disease.

Febrile convulsions ➔ p. 875

Red	**Amber**	**Green**
High-risk symptoms/signs	***Intermediate-risk symptoms/signs***	***Low-risk symptoms/signs***
Appears ill Colour—pale, mottled, ashen, or blue No response to social cues Does not wake or if roused does not stay awake Weak, high-pitched, or continuous cry Grunting Respiratory rate >60/min Moderate/severe chest in-drawing ↓ skin turgor High temperature of ≥38°C aged <3mo (though some vaccinations induce fever in children <3 mo) Non-blanching rash Bulging fontanelle Neck stiffness Status epilepticus Focal neurological signs/seizures	Pallor ↓ response to social cues Wakes only with excessive stimulation ↓ activity No smile Nasal flaring ↑ respiratory rate (>50/min aged 6–12mo; >40/min aged >12mo) O_2 saturation ≤95% in air Crackles in chest Tachycardia (>160bpm age <12mo; >150bpm age 12–24mo; >140bpm age 2–5y) Capillary return ≥3sec High temperature ≥39°C age 3–6mo Rigors Dry mucous membranes Poor feeding in infants ↓ urine output Swelling of a limb/joint Non-weight bearing/ not using an extremity	Normal colour of skin/lips/tongue Responds normally to social cues Content/smiles Stays awake or awakens quickly Strong normal cry or not crying Normal skin/eye turgor Moist mucous membranes None of the amber/red symptoms or signs

Figure 23.2 Traffic light system for assessment of children <5y with fever[N]

⚠ **Acute illness in non-febrile children** Sick children do not have to have a fever; refer for immediate or same-day paediatric review if:

- Any symptoms or signs in the Red column of Figure 23.2
- Persistent vomiting—more than half of the previous 3 feeds or bile stained
- Frank blood in the stools or urine
- History suggestive of apnoeic episodes

Further information

NICE (2013, updated 2017) Fever in under 5s: assessment and initial management. 🖱 www.nice.org.uk/guidance/cg160
Spotting the sick child 🖱 www.spottingthesickchild.com

Childhood infection

Table 23.8 A–Z of childhood infection

Infection	Page	Infection	Page
Chickenpox	➔ p. 629	Malaria	➔ p. 624
Conjunctivitis	➔ p. 944	Measles	➔ p. 629
Croup	➔ p. 915	Molluscum contagiosum	➔ p. 609
Diphtheria	➔ p. 633	Mumps	➔ p. 629
Epiglottitis	➔ p. 915	Otitis media	➔ p. 924
Erythema infectiosum	➔ p. 629	Polio	➔ p. 551
Gastroenteritis	➔ p. 348	Roseola infantum	➔ p. 629
Glandular fever	➔ p. 913	Rubella	➔ p. 629
Hand, foot, and mouth	➔ p. 629	Scabies	➔ p. 613
Head lice	➔ p. 612	Scarlet fever	➔ p. 630
Hepatitis A	➔ p. 394	Shingles	➔ p. 628
Hepatitis B/C	➔ p. 718	Sinusitis	➔ p. 920
Herpes (skin)	➔ p. 608	Skin infection	➔ p. 606
HIV	➔ p. 722	Tuberculosis (TB)	➔ p. 296
Impetigo	➔ p. 606	Tonsillitis	➔ p. 912
Influenza	➔ p. 292	UTI in childhood	➔ p. 856
Kawasaki's disease	➔ p. 501	Warts and verrucas	➔ p. 608
Lyme disease	➔ p. 571	Whooping cough	➔ p. 298

Viral upper respiratory tract infection (URTI) Common—each child has >5 URTIs each year. Presents with coryza, runny eyes, malaise ± mild pyrexia, and/or non-specific maculopapular rash. Examine to exclude tonsillitis and otitis media. If pyrexia but no other symptoms/signs, check urine to exclude UTI. Most viral URTIs settle within a few days.

Childhood pneumonia May be viral, bacterial (e.g. pneumococcal, *Haemophilus influenzae*, staphylococcal), or atypical (e.g. mycoplasma). Presents with ≥1 of:

- Fever—recurrent or persistent >38.5°C
- Cough
- Chest and/or abdominal pain
- Tachypnoea, recession, or other signs of respiratory difficulty
- Crepitations, ↓ breath sounds, ± bronchial breathing

Assessing severity Severe if:

- Any 'Red' features from Figure 23.2 (age <5y), ➔ p. 853, Figure 29.11 (age ≥12y), ➔ p. 1057 or Figure 29.12 (age 5–12y), ➔ p. 1058
- Peripheral O_2 saturation <92% and/or cyanosis
- Tachypnoea >70 breaths/min aged <1y; >50 breaths/min aged >1y
- Difficulty breathing/grunting
- Dehydrated (or not feeding if <1y)
- Not responding to antibiotics
- Family are unable to manage

⚠ If any 'severe' features, admit immediately to paediatrics. If any 'Amber' features, e.g., peripheral O_2 saturation of <95% or capillary refill time ≥3s, consider admission or arrange early review.

Management in the community

- Provide advice about management of fever and hydration

- Well children with mild symptoms of LRTI may not require antibiotics
- If prescribing antibiotics, amoxicillin is first-line; consider a macrolide if penicillin allergic, *Mycoplasma* or *Chlamydia* is suspected as the cause of the infection, or in addition to amoxicillin if first-line treatment is ineffective. Use co-amoxiclav as first-line if pneumonia associated with influenza
- Advise parents to seek further medical review if no better in <48h (sooner if any 'Amber' features) or worse in the interim

Prevention Pneumococcal vaccination is part of the routine childhood vaccination programme and is given at 2, 4, and 12mo (➔ p. 619).

Recurrent chest infection Consider further investigation and/or referral to look for an underlying cause if a child has a history of 2 probable chest infections. Possible underlying causes include:

- Asthma
- Post-infective bronchiectasis
- Congenital heart/lung disease
- TB
- Sickle cell disease
- 💣 Right middle lobe syndrome
- Cystic fibrosis
- Foreign body
- Oropharyngeal aspiration, e.g. due to reflux
- Immune disorders, e.g. HIV, hypogammaglobulinaemia, leukaemia

Bronchiolitis[N] Common lower respiratory tract infection in infants aged <1y. Occurs in epidemics—usually in winter. Mostly caused by respiratory syncytial virus (RSV). Infants at ↑ risk of severe disease include: premature babies; very young babies (<12wk old); and children with underlying lung disease, congenital heart disease, neuromuscular disease, or immunosuppression.

Prevention Palivizumab (a monoclonal antibody) ↓ RSV infection rate and severity. It is consultant initiated and given IM to premature babies, and those at ↑ risk from RSV infection due to other co-morbidities, before the RSV season and then monthly until the end of the season.

Presentation Coryza (1–3d) followed by persistent cough, rapid breathing ± feeding difficulty. May present with apnoea. *Examination*: tachypnoea; recession; widespread crepitations/wheeze; fever (30%—usually <39°C—if higher consider pneumonia).

Management Check peripheral O_2 saturation if available.

- **Admit as paediatric emergency** If any 'Red' features in Figure 23.2 (➔ p. 853), lethargy/exhaustion, looks seriously unwell, marked intercostal recession, grunting, respiratory rate >70 breaths/min, cyanosis, peripheral O_2 saturation <92% or apnoeic episode(s) (reported or observed)
- **Consider same-day paediatric referral** If respiratory rate >60 breaths/min, taking <½–¾ usual feeds, or dehydrated. Have a lower threshold for admission if poor social circumstances, long distance from hospital, and/or if high risk infant
- **Home management** Is safe if feeding well, no/mild recession. Advise parents not to smoke in the home as smoking ↑ risk of severe symptoms, how to recognize worsening symptoms, and how to call for help

Prognosis Most recover in <14d; up to 50% wheeze with subsequent URTIs.

Further information

NICE (2015) Bronchiolitis in children: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng9

Urinary tract infection in childhood

10% girls and 3.5% boys have a urinary tract infection (UTI) in childhood—the majority in the first year of life. Among neonates, boys have more infections than girls. In all other age groups, ♀:♂ ≈10:1. 80% of infections are due to *Escherichia coli*.

Risk factors

- Poor urine flow
- History suggesting/confirmed past UTI
- Recurrent fever of unknown origin
- Antenatally diagnosed renal abnormality
- FH of vesico-ureteric reflux (VUR) or renal disease
- Dysfunctional voiding
- Constipation
- Enlarged bladder
- Abdominal mass
- Spinal lesion
- Poor growth
- ↑ blood pressure

Consequences 5–15% develop renal scarring <2y after first infection. Infections causing renal scarring are associated with adult pyelonephritis, ↑ BP, impaired renal function, and renal failure. Prognosis is worst for children with recurrent infection, VUR, and scarring at first presentation.

Clinical presentation Depends on age and site of infection:

- **Infants and toddlers** Usually non-specific with vomiting, irritability, fever, abdominal pain, failure to thrive, and (in infants) prolonged jaundice
- **Older children** Dysuria, ↑ frequency, abdominal pain, haematuria, enuresis
- **Site** Fever >38°C and/or loin pain/tenderness suggests upper UTI/acute pyelonephritis

Babies <3mo with suspected UTI should be referred to paediatrics for immediate assessment as IV antibiotics are recommended.

Management of children aged >3mo^{N}

Presentation with fever Suspect UTI in any child with fever >38°C with no obvious cause. Use the traffic light system (➔ p. 853) to guide management. If any 'Red' features, admit/refer for emergency assessment.

Urine testing Check urine dipstick if signs/symptoms of UTI, unexplained fever, or if a child fails to recover from a fever presumed due to another cause. A clean catch specimen is best; otherwise use a special bag/pad. Depending on the result, consider sending a urine sample to the laboratory for M,C&S and/or starting antibiotics—Table 23.9.

Antibiotics Prescribe for 3d for lower UTI and 7–10d for upper UTI. Choice of antibiotic depends on local guidelines, but first line is usually trimethoprim or nitrofurantoin. Review after 24–48h if not improving, and send a urine sample for M,C&S if a sample has not already been sent.

Interpretation of microscopy results

- All children with significant bacteriuria should be treated with antibiotics
- If pyuria but no bacteriuria, start antibiotics if clinical features of UTI
- If no pyuria and no bacteriuria, the child does not have a UTI

Follow-up Do not check M,C&S to confirm eradication. Only start prophylactic antibiotics if >1 UTI. Treat any constipation. Advise to drink plenty of fluids. Consider further investigations.

Table 23.9 Management based on urine dipstick test results[N]

Urine dipstick result		Management
Leukocyte esterase	*Nitrite*	
Positive	Positive	Start antibiotics; send urine for M,C&S if: • Aged <3y • Past history of UTI • Suspected upper UTI/pyelonephritis • Child has any 'Amber' features and is being managed at home (age <5y -Figure 23.2, ➔ p. 853; age 5–12y—Figure 29.12, ➔ p. 1058; age ≥12y—Figure 29.11, ➔ p. 1057) • Not improving after 24–48h on antibiotics
Negative	Positive	Start antibiotics; always send urine for M,C&S
Positive	Negative	Send urine for M,C&S; only start antibiotics if: • Aged <3y • Strong clinical evidence for UTI • UTI is proven on laboratory testing
Negative	Negative	Do not start antibiotics; look for other causes of symptoms Send urine for M,C&S if the child: • Has clinical features of UTI despite dipstick results • Appears unwell with no apparent cause • Has a past history of urine infection

Further investigation ❗ Arrangements vary in the UK. Most GPs have direct access to USS. Other investigations may require referral to paediatrics.

Children <6mo responding to antibiotics in <48h Arrange USS ± micturating cystourethrogram (MCUG) depending on USS findings.

Children of any age with atypical infection Defined as any of:
- Child very unwell (any 'Amber'/'Red' features from sepsis tables)
- Response to antibiotics took >48h
- ↑ creatinine
- Poor urine flow
- Non-*E. coli* infection
- Abdominal/bladder mass

Arrange urgent USS in all cases, plus:
- DMSA scan if <3y of age, and
- MCUG if <6mo, or if <3y and dilation on USS, poor urine flow, non-*E. coli* infection, or family history of vesico-ureteric reflux

Children of any age with recurrent infection Defined as:
- ≥2 upper UTI
- ≥3 lower UTI, or
- 1 upper UTI + ≥1 lower UTI

Arrange USS (urgent if <6mo old) + DMSA scan ± MCUG (if aged <6mo).

Balanitis ➔ p. 438 **Epididymo-orchitis** ➔ p. 440

Horseshoe kidney, ectopic kidney, double ureter Common urinary tract malformations. Usually do not affect kidney function per se but predispose to UTI. Recurrent infections may eventually cause renal damage.

Further information

NICE (2007, updated 2018) Urinary tract infection in under 16s: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg54

Congenital heart disease

Common, affecting ~6 in 1000 live births. Congenital heart disease is the major cause of heart disease in children (Table 23.10).

Detection

Antenatal screening ~50% of severe congenital heart disease is detected *in utero* by USS. If detected during the routine 8–14wk or 18–21wk scan, amniocentesis is routinely offered to screen for Down's syndrome (~1 in 20 have heart disease—especially PDA, ASD, and/or VSD) and other chromosomal abnormalities.

Clinical examination Neonatal examination detects <50% of cardiac malformations not detected antenatally. Pulse oximetry is being considered as a way of ↑ detection rates. The rest are detected if a murmur is found incidentally when examining the child for another reason or when the child becomes symptomatic.

Presentation

Murmur on examination Murmurs are a common finding in childhood particularly when examining a febrile child. The majority are not associated with heart disease—so-called '*innocent*' murmurs. *Features*:

- Asymptomatic
- Soft, systolic murmur—may vary with position and does not radiate
- Normal 2nd heart sound
- No other associated signs of heart disease (normal pulses, no thrill)

❗ Unless the child is febrile when the murmur is heard, and it disappears once afebrile, refer all children with murmurs for echo or paediatric evaluation.

Once a murmur is known to be 'innocent', explain what that means to the parents—otherwise there may be unnecessary ongoing anxiety.

Murmurs associated with congenital heart disease include:

- **Ventriculoseptal defect (VSD)** Harsh pansystolic murmur with splitting of the 2nd heart sound
- **Atrioseptal defect (ASD)** Systolic murmur in the pulmonary area with fixed splitting of the 2nd heart sound
- **Patent ductus arteriosus (PDA)** Loud, continual 'machinery' murmur
- **Aortic stenosis** Ejection systolic murmur at the apex and left sternal edge with a soft and delayed 2nd heart sound. Slow rising pulse, ↓ BP. Rarely dizziness, faintness, or loss of consciousness on exertion
- **Pulmonary stenosis** Ejection systolic murmur with ejection click
- **Coarctation of the aorta** Ejection systolic murmur over the left side and back; absent/delayed femoral pulses and ↑ BP just in upper limbs

Cyanosis

- **<48h old** Likely to be due to transposition of the great arteries or severe pulmonary stenosis
- **Later presentation** Mostly due to *tetralogy of Fallot* (Table 23.10)

Table 23.10 Congenital cardiac abnormalities

Condition	Features
ASD	➲ p. 252
Coarctation of the aorta	➲ p. 252
Tetralogy of Fallot	Large VSD and pulmonary stenosis In the newborn period may present with a murmur Progressive cyanosis then develops over the next weeks/years ± ↓ exercise tolerance ± squatting after exercise Treatment is surgical
Hypoplastic left heart	Left ventricle ± mitral valve, aortic valve, and aortic arch are underdeveloped Presents within the 1st few days of life with heart failure Treatment is surgical or with heart transplant
Patent ductus arteriosus (PDA)	The ductus arteriosus fails to close after birth ♀ > ♂. Associated with prematurity Symptoms depend on the size of the shunt. Presents with murmur ± faltering growth ± heart failure Treatment is usually surgical closure
Transposition of the great arteries	The aorta arises from the right ventricle and the pulmonary artery from the left Progressive cyanosis develops within a few hours of birth Treatment is surgical
Valve disease	➲ p. 250
VSD	➲ p. 252

Heart failure

- Breathlessness particularly when crying/feeding
- Failure to thrive
- Sweating
- Fast respiratory and pulse rates
- Heart enlargement
- Liver enlargement
- Weight ↑ due to fluid retention

Causes of heart failure in the first week of life include:

- Left outflow obstruction
- Severe aortic stenosis
- Coarctation of the aorta
- Hypoplastic left heart

Later causes:

- Large VSD
- PDA
- Ostium primum ASD

Management In all cases, if new congenital heart disease is suspected, refer for specialist paediatric or cardiology opinion. Specialist treatment of valve lesions depends on the gradient measured across the valve. Most other congenital cardiac lesions (except some VSDs and ASDs) require surgery—staged for complex lesions.

Information and support for parents

Children's Heart Federation ☎ 0808 808 5000 🖱 www.chfed.org.uk

Diagnosis of asthma in children

Childhood asthma affects ~5% of children in the UK. Peak age of onset is 5y. In the UK ~20 children <14y old still die from asthma each year.

💣 There are two competing national asthma guidelines in the UK which differ significantly from each other. The guidance in this section is based on BTS/SIGN guidance; a link to the parallel NICE guidance is provided.

Diagnosis Use a structured clinical assessment to determine if there is a high/intermediate/low probability of asthma, including history, examination, and reference to previous assessments and tests.

High probability of asthma

- Recurrent, episodic asthma symptoms (>1 of wheeze, breathlessness, chest tightness), which may occur in response to, or are worse after, exercise, other triggers (e.g. pets, cold/damp air), or with emotions/laughter
- Diurnal variability of symptoms
- Expiratory wheeze on auscultation
- Documented variable airflow obstruction, e.g. PEFR, spirometry
- Personal or family history of atopy
- Absence of features to suggest an alternative diagnosis

Intermediate probability of asthma Some, but not all, the features of asthma, or poor response to treatment.

Low probability of asthma Absence of typical asthma features or presence of symptoms suggesting an alternative diagnosis—see Table 23.11, ➔ p. 865.

❗ Normal spirometry/PEFR testing, if performed when the child is asymptomatic, does not exclude asthma.

Clinical features that ↓ the likelihood of asthma include

- Symptoms with colds only and no interval symptoms—virus-associated wheeze affects up to 20% of children at some point
- Isolated cough in the absence of wheeze or difficulty breathing
- History of moist cough
- Prominent dizziness, light-headedness, peripheral tingling
- Repeatedly normal physical examination of chest when symptomatic
- Normal PEFR or spirometry when symptomatic
- No response to a trial of asthma therapy

If high probability of asthma Start a trial of treatment (➔ p. 862). Review and assess response. If +ve response, a diagnosis of asthma can be made. 💣 NICE recommends testing with spirometry, FeNO testing, and/or serial PEFR for all children >5y (or when old enough to test).

If intermediate probability of asthma Arrange:

- **Spirometry** Usually possible if aged >5y. Use lower limit of normal for age for FEV$_1$/FVC to detect airways obstruction (BTS) (NICE states <70%)
- **Reversibility** If evidence of airway obstruction, assess change in FEV$_1$ in response to bronchodilator and/or response to trial of treatment with inhaled steroids. ↑ in FEV$_1$ of >12% from baseline and/or beneficial treatment trial supports a diagnosis of asthma

- If unable to perform spirometry reliably, consider:
  - Watchful waiting with review if mild, intermittent wheeze and/or symptoms that occur only with viral URTIs
  - Monitored initiation of treatment for 6–8wk, continue if response. If no response, consider further investigation as for low probability. If it is unclear if a child has improved, try withdrawing the treatment
- If no evidence of airways obstruction, arrange FeNO test or challenge test for bronchial hyper-responsiveness (e.g. exercise challenge in those with symptoms brought on by exercise). Consider specialist referral

💣 NICE recommends an initial 8wk trial of inhaled steroids for all children <5y with suspected, confirmed, or uncontrolled asthma.

If low probability of asthma Consider more detailed investigation for asthma and alternative diagnoses and/or specialist referral.

❗ Do a CXR if severe disease or clinical suggestion of another condition.

Reasons for referral *E* = Emergency; *U* = Urgent; *S* = Soon; *R* = Routine. ❗ This is only a rough guide; urgency of referral depends on the clinical state of the child:

- Severe exacerbation of asthma or severe URTI—*E*
- Unexpected clinical findings, e.g. focal signs, abnormal voice or cry, dysphagia, inspiratory stridor—*E/U*
- Persistent wet or productive cough—*U/S*
- Faltering growth (failure to thrive)—*U/S*
- Diagnosis unclear or in doubt—*U/S/R*
- Failure to respond to conventional treatment (particularly inhaled corticosteroids >400 micrograms/d or frequent use of steroid tablets)—*S*
- Excessive vomiting or posseting—*S/R*
- Symptoms present from birth or perinatal lung problem—*S/R*
- FH of unusual chest disease—*R*
- Nasal polyps—*R*
- Parental anxiety or need for reassurance—*R*

Differential diagnosis See Table 23.11, ➔ p. 865.

Prognostic factors

- The earlier the onset of wheeze, the better the prognosis—most children presenting at <2y become asymptomatic by mid childhood
- Male gender—risk factor for asthma in pre-pubertal children—but boys are more likely to 'grow out' of asthma during adolescence
- Coexistent/FH of atopy—risk factor for persistence of wheeze
- Frequent/severe episodes of wheezing in childhood—associated with recurrent wheeze that persists into adolescence; persistent abnormal lung function is associated with asthma in adult life

Further information

BTS/SIGN (2019) British guideline on the management of asthma 🖱 www.sign.ac.uk/sign-158-british-guideline-on-the-management-of-asthma.html

NICE (2017) Asthma: diagnosis, monitoring and chronic asthma management 🖱 www.nice.org.uk/guidance/ng80

Information and support for parents and patients

Asthma UK ☎ 0800 1216244 🖱 www.asthma.org.uk

Management of asthma in children

Symptoms/signs of a significant asthma attack in a child >2y

- Peripheral O_2 saturation <92%
- PEFR 33–50% best/predicted (>5y)
- Too breathless to talk
- ↑ heart rate (>140bpm age 2–5y; >125 bpm age >5y)
- ↑ respiratory rate (>40 breaths/min age 2–5y; >30 breaths/min age >5y)
- Use of accessory neck muscles

Life-threatening signs in a child >2y Peripheral O_2 saturation <92% plus ≥1 of:

- PEFR <33% best/predicted (>5y)
- Silent chest (inaudible wheeze)
- Poor respiratory effort
- Agitation
- Cyanosis
- Altered consciousness

Symptoms/signs of a severe asthma attack in children <2y

Life-threatening features

- Peripheral O_2 saturation of <92%
- Marked respiratory distress
- Cyanosis
- Too breathless to feed
- Episodes of apnoea
- Poor respiratory effort
- Bradycardia

Management of an acute asthma attack ➲ p. 1075

Aim of treatment To control the disease to:

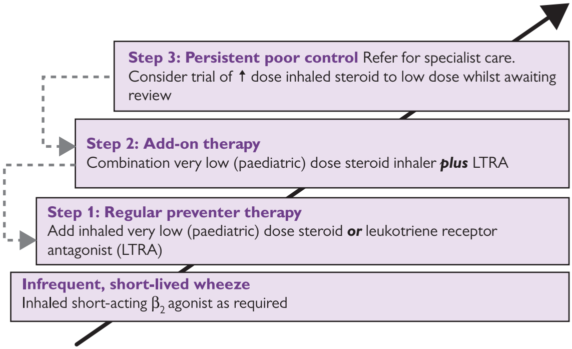
- Minimize symptoms and impact on lifestyle (e.g. absence from school; limitations to physical ability; night-time waking)
- Minimize the need for reliever medication
- Prevent severe attacks/exacerbations
- Minimize medication side effects
- Maintain normal lung function

GP services and self-management ➲ p. 280. For children, ensure that the following are checked and recorded at least annually:

- Symptom score, e.g. Children's Asthma Control Test (from 4–11y), Asthma Control Questionnaire (>5y)
- Exacerbations, oral steroid use, and time off school/nursery due to asthma since last check
- Inhaler technique and medication adherence (prescription frequency)
- Written personalized asthma self-management action plan
- Exposure to tobacco smoke (e.g. parental smoking)
- Growth—height and weight centile

Drug therapy Use a stepwise approach (➲ p. 863). Start at a step appropriate to the initial severity of symptoms. Aim to achieve control of the condition and then ↓ by stepping down.

Exacerbations Treat exacerbations early. A rescue course of prednisolone (30–40mg/d if aged >5y or 20mg/d if aged 2–5y) for 3–14d may be needed at any step and any time. Alternatively, leukotriene antagonists can be used for children with episodic asthma—start at the onset of asthma/coryzal symptoms and continue for 7d.



Step 3: Persistent poor control Refer for specialist care. Consider trial of ↑ dose inhaled steroid to low dose whilst awaiting review

Step 2: Add-on therapy
Combination very low (paediatric) dose steroid inhaler ***plus*** LTRA

Step 1: Regular preventer therapy
Add inhaled very low (paediatric) dose steroid ***or*** leukotriene receptor antagonist (LTRA)

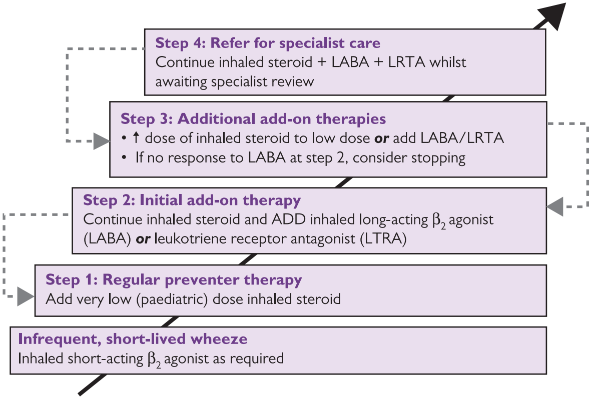
Infrequent, short-lived wheeze
Inhaled short-acting β_2 agonist as required

Consider moving up a step if using ≥3 doses of short-acting β_2 agonist per week.

NICE recommends an initial 8wk trial of inhaled steroids for all children <5y with suspected, confirmed, or uncontrolled asthma.

Figure 23.3 Stepwise management of asthma in children aged <5y

Source: data from British Thoracic Society and Scottish Intercollegiate Guidelines Network, *SIGN 158: British guideline on the management of asthma* (revised 2019) https://www.sign.ac.uk/assets/sign158.pdf



Step 4: Refer for specialist care
Continue inhaled steroid + LABA + LRTA whilst awaiting specialist review

Step 3: Additional add-on therapies
- ↑ dose of inhaled steroid to low dose ***or*** add LABA/LRTA
- If no response to LABA at step 2, consider stopping

Step 2: Initial add-on therapy
Continue inhaled steroid and ADD inhaled long-acting β_2 agonist (LABA) ***or*** leukotriene receptor antagonist (LTRA)

Step 1: Regular preventer therapy
Add very low (paediatric) dose inhaled steroid

Infrequent, short-lived wheeze
Inhaled short-acting β_2 agonist as required

Consider moving up a step if using ≥3 doses of short-acting β_2 agonist per week

NICE recommends trying leukotriene receptor antagonists (LRTA) rather than LABA as initial add-on therapy. If ineffective, NICE recommends stopping the LRTA and adding a LABA in a combined LABA/inhaled steroid inhaler.

Figure 23.4 Stepwise management of asthma in children aged 5–12y

Source: data from British Thoracic Society and Scottish Intercollegiate Guidelines Network, *SIGN 158: British guideline on the management of asthma* (revised 2019) https://www.sign.ac.uk/assets/sign158.pdf

Short-acting β_2 agonists (➔ p. 276) (e.g. salbutamol). Bronchodilator. Works more quickly and with fewer side effects than alternatives. PRN dosing is at least as effective as regular dosing. Using ≥1 canister/mo is a marker of poorly controlled asthma.

Inhaled corticosteroids (➔ p. 276) Most effective asthma preventer. May be beneficial even for children with mild asthma. Consider if:

- Exacerbations needing oral corticosteroids in the last 2y (age >5y)
- Using inhaled β_2-agonists ≥3×/wk
- Symptomatic ≥3×/wk or ≥1 night/wk

Oral steroids ➔ p. 276

Add-on therapy Before initiating a new drug, check compliance, inhaler technique, and eliminate trigger factors.

- Inhaled long-acting β_2 agonists (➔ p. 276) (e.g. salmeterol). First-choice add-on therapy for children aged >5y to improve lung function and symptoms. Do not use without inhaled steroid—use of combination inhalers is recommended. Stop if of no benefit
- Leukotriene receptor antagonists (e.g. montelukast). May ↓ symptoms and exacerbations and ↑ lung function. First-choice add-on therapy for children <5y
- Theophylline or β_2-agonist slow-release tablets May ↓ symptoms and ↑ lung function but side effects are common

Stepping down Review and consider stepping down at intervals of ≥3mo. Maintain on the lowest dose of inhaled steroid controlling symptoms. When reducing steroids, cut dose by 25–50% each time. Children with milder asthma and a clear seasonal pattern can step down more quickly in their good season.

Selection of inhaler device If possible use a metred-dose inhaler. Inadequate technique may be mistaken for drug failure. Emphasize patients must inhale slowly and hold their breath for 10sec after inhalation. Demonstrate inhaler technique before prescribing and check at follow-ups. Spacers are useful for children who find activation difficult and essential for children <5y and when higher doses of steroids are used. Dry powder or breath-activated inhalers are an alternative for older children.

Expected PEFR in children ➔ p. 274

Allergen avoidance ➔ p. 281

Complementary therapy ➔ p. 282

Asthma in adolescents Common but may be under-diagnosed due to under-reporting of symptoms, so specifically ask about symptoms:

- Breathlessness/wheezing with exercise is common; if due to asthma, it should respond to pre-treatment with a β_2-agonist
- Depression/anxiety may make asthma symptoms more prominent
- Advice about risks of smoking/passive smoking is very important
- Inhaler preference is important in ensuring good treatment adherence
- Complementary or alternative medicine use is common and may be linked with adherence to prescribed medication
- Discuss career choice and highlight occupations that may ↑ symptoms

Table 23.11 Differential diagnosis of wheezing in children[G]

Clinical clue	Possible diagnosis
Perinatal and family history	
Symptoms present from birth or perinatal lung problem	Cystic fibrosis; chronic lung disease of prematurity; ciliary dyskinesia; developmental lung anomaly
Family history of unusual chest disease	Cystic fibrosis; neuromuscular disorder
Severe upper respiratory tract disease	Defect of host defence; ciliary dyskinesia
Symptoms and signs	
Persistent moist cough	Cystic fibrosis; bronchiectasis; protracted bacterial bronchitis; recurrent aspiration; host defence disorder; ciliary dyskinesia
Excessive vomiting	Gastro-oesophageal reflux (with or without aspiration)
Paroxysmal coughing bouts leading to vomiting	Pertussis
Dysphagia	Swallowing problems (with or without aspiration)
Breathlessness with light-headedness and peripheral tingling	Dysfunctional breathing; panic attacks
Inspiratory stridor	Tracheal or laryngeal disorder
Abnormal voice or cry	Laryngeal problem
Focal signs in chest	Developmental anomaly; post-infective syndrome; bronchiectasis; tuberculosis
Finger clubbing	Cystic fibrosis; bronchiectasis
Failure to thrive	Cystic fibrosis; host defence disorder; gastro-oesophageal reflux
Investigations	
Focal or persistent radiological changes	Developmental lung anomaly; cystic fibrosis; postinfective disorder; recurrent aspiration; inhaled foreign body; bronchiectasis; tuberculosis

Further information

BTS/SIGN (2019) British guideline on the management of asthma. 🖰 www.sign.ac.uk/sign-158-british-guideline-on-the-management-of-asthma.html

NICE (2017) Asthma: diagnosis, monitoring and chronic asthma management. 🖰 www.nice.org.uk/guidance/ng80

Information and support for parents and patients

Asthma UK ☎ 0800 1216244 🖰 www.asthma.org.uk

Constipation and malabsorption

Constipation[N] Frequent complaint among all age groups of children. Take a careful history—diagnose constipation if ≥2 of:

- Abnormal stool pattern:
  - <3 stool per week (except breastfed babies >6wk)
  - Hard, large, or rabbit-dropping stool
  - Overflow soiling (children >1y)
- Symptoms associated with defecation:
  - Distress on stooling or anal pain
  - Bleeding associated with hard stool
  - Straining
  - Poor appetite or abdominal pain that improves after stool passed
  - Posture indicating retaining stool (straight legs, back arch, tiptoe)
- History of previous constipation or anal fissure

Serious underlying causes

- Hirschsprung's disease
- Coeliac disease
- Hypothyroidism
- Anorectal abnormalities
- Neurological conditions
- Abdominal tumours

Management Refer for specialist assessment if any red flag symptoms/signs suggesting a serious underlying cause:

- Delay in passing meconium or constipation since birth
- Abnormal appearance, position, or patency of the anus
- Ribbon-like stool
- New leg weakness, deformity (e.g. talipes), or neuromuscular signs
- Asymmetrical gluteal muscles, sacral naevus, sinus, or pit, or scoliosis
- Abdominal distension with vomiting or gross distension
- If there is faltering growth, check coeliac serology and TFTs

Treatment of idiopathic constipation Diagnose if serious underlying causes have been excluded. Advise about balanced diet and adequate fluid intake. Use a macrogol as first-line laxative adjusting dose according to response. Add a stimulant laxative if ineffective. If macrogols are not tolerated use a stimulant ± alternative softener. Continue medication for several weeks after regular bowel habit has been established. Refer for specialist assessment if not responding to treatment in ≤3mo (≤1mo if <1y old).

Diarrhoea and vomiting ➔ p. 346

Malabsorption In all cases refer for investigation and treatment of the cause. Usually presents with:

- Chronic diarrhoea
- Faltering growth or weight ↓
- Steatorrhoea, and/or
- Iron or other nutrient deficiency

Causes in children

- **Common** Cow's milk intolerance (cow's milk protein allergy, or lactose intolerance); coeliac disease
- **Rarer** Cystic fibrosis; chronic infection (e.g. giardiasis); inflammatory bowel disease

Cow's milk protein allergy^{N} Affects ~3% of children. Mainly occurs in bottle-fed infants <6mo old but affects 0.5% of purely breastfed babies as cow's milk protein from the mother's diet is secreted in breast milk.

Presentation May be FH. Often affects >1 body system:

- Gastrointestinal symptoms (50–60%) Diarrhoea (occasionally with blood); colic; less commonly constipation
- Skin symptoms (50–70%) Urticaria; eczema
- Respiratory and other symptoms (20–30%) Wheeze; rhinitis; conjunctivitis

Diagnosis Often clinical—try withdrawing cow's milk. If this resolves symptoms, and symptoms return with reintroduction, cow's milk protein allergy is likely. Skin prick/RAST tests have high false +ve and –ve results.

Treatment

- Eliminate cow's milk—usually by replacing cow's milk formula with hydrolysed protein milk formula (e.g. Nutramigen®)
- 10% of babies are also intolerant to hydrolysed protein formula and require amino acid formula (e.g. Neocate®)
- Advise parents that solids should be dairy free
- Most children grow out of cow's milk protein intolerance and can be challenged with foods containing milk from 12mo

Lactose intolerance Rare in infancy; more common in childhood.

Presentation in infancy Severe symptoms of abdominal distension, diarrhoea (explosive and watery), ± vomiting and faltering growth. Stools test +ve for reducing sugars. Treatment is with a lactose-free diet.

Presentation in later childhood (>2y) Due to lactase deficiency.

- Common—incidence is very variable depending on place of origin—but for people of European origin ~10–15% (higher if of Asian origin)
- Symptoms tend to be milder with abdominal pain and/or distension ± diarrhoea and/or vomiting appearing at any time from 2y of age to adulthood. Symptoms improve when milk products are removed from the diet. Tolerance of milk products is variable—cheese and yoghurt are better tolerated as lactose is hydrolysed by bacteria

❗ Soya milk does not contain lactose. Milk from other animals (e.g. goat) does contain lactose. Soya milk contains phyto-oestrogens and has high glucose content, do not use if <6mo of age or long term except on specialist advice.

Cow's milk intolerance following gastroenteritis Temporary cow's milk intolerance is common after a bout of gastroenteritis in children and can result in continuing diarrhoea (>2wk). Try excluding cow's milk from the diet. If symptoms improve, wait until all symptoms have gone and then reintroduce cow's milk. If symptoms do not improve, refer for further investigation.

Further information

NICE (2010, updated 2017) Constipation in children and young people: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg99

NICE (2011) Food allergy in under 19s: assessment and diagnosis. 🖱 www.nice.org.uk/guidance/cg116

Gut atresia, hernias, and intussusception

Oesophageal atresia ➔ p. 357

Duodenal atresia Usually associated with other abnormalities—particularly Down's syndrome. If not detected on antenatal USS, presents postnatally with bile-stained vomiting. AXR reveals a 'double bubble' with air in the stomach and first part of duodenum but none beyond. Requires surgical correction.

Anorectal atresia (imperforate anus) 1 in 4000 live births. Usually the baby fails to pass meconium and no anus is visible, or anus may be stenosed or in wrong place. There is often a fistula to the urethra (boys) or vagina (girls). Treatment is surgical and may require colostomy for high lesions. In the period after surgery, anal dilation is vital to prevent stricture. It requires use of graded dilators by the baby's parents for several months. Faecal incontinence may be a problem but can usually be managed using a combination of dietary manipulation, enemas, and drug treatment.

Umbilical hernia Common. Presents as a bulge at the umbilicus. Painless and reducible, but more prominent when the baby cries. Due to a defect in the umbilical ring when the cord separates. More common in people of black ethnic origin, premature and low birthweight babies, and associated with certain syndromes (e.g. trisomy 13 and 18 or 21). Usually resolves spontaneously. Strangulation is rare. Refer for surgery if an umbilical hernia persists until >4y of age.

Inguinal hernia More common in premature, low birthweight, and ♂ babies.

- Non-acute History of intermittent groin ± scrotal swelling—the spermatic cord may be thickened on the affected side. Refer to paediatric surgery for repair (herniotomy). Risk of strangulation is greatest in first year, particularly in premature babies, and ↓ with age
- Acute Sudden appearance of an irreducible groin or scrotal swelling—necessitates emergency admission for reduction and repair

Diaphragmatic hernia *Incidence*: 1 in 2500 live births. A defect in one hemidiaphragm allows the bowel to herniate into the chest cavity → pulmonary hypoplasia *in utero* or lung compression postnatally. Detected antenatally on USS or postnatally when the child develops respiratory distress soon after birth—CXR confirms diagnosis. Corrective surgery is associated with high mortality but once successfully repaired the child usually has no further difficulties.

Exomphalos and gastroschisis

- Exomphalos Complete return of the gut into the abdominal cavity fails to occur during intrauterine life. At birth there is a swelling at the umbilicus, consisting of gut covered by a membrane
- Gastroschisis There is a defect in the abdominal wall through which exposed gut prolapses

Exomphalos and gastroschisis are usually detected antenatally at routine USS. Delivery then takes place at a specialist centre where surgical repair can be undertaken soon after birth. Once repaired prognosis is good.

Intussusception The invagination of one part of the bowel into the lumen of the immediately adjoining bowel. It is the most common cause of intestinal obstruction in young children and usually occurs in previously healthy children. *Incidence*: 2 in 1000 live births. *Peak age*: 5–18mo; ♂:♀ ≈2:1.

Associations

- Seasonal variation suggests an underlying viral cause—rota- and adenoviruses have both been implicated
- Intestinal polyps
- Meckel's diverticulum
- Henoch–Schönlein purpura

Types

- **Ileo-ileal** Ileum invaginates into adjacent ileum
- **Ileo-colic** Most common type—an ileo-ileal intussusception extends through the ileo-caecal valve
- **Ileo-caecal** The apex of the intussusception is the ileo-caecal valve
- **Colo-colic** Colon invaginates into adjacent colon—may be secondary to bowel tumour

Presentation Very variable. Always have a high index of suspicion.

- Abdominal colic—paroxysms of pain during which the child draws up his/her legs—the child often screams with the pain and becomes pale. Episodes usually are 10–15min apart and last 2–3min but become more frequent with time
- Vomiting—early symptom
- Rectal bleeding—passage of blood ('redcurrant jelly stool') or slime per rectum is a late sign
- Sausage-shaped mass in the abdomen—usually in the right upper quadrant—though not always present

⚠ The child becomes rapidly worse if not treated early, becoming toxic and developing obstruction with distended abdomen ± feculent vomiting.

Differential diagnosis Other causes of bowel obstruction; gastroenteritis; constipation; haemolytic uraemic syndrome.

Management Admit as an acute paediatric or paediatric surgery emergency (depending on local pathways). Untreated intussusception is usually fatal. Treatment is with reduction by air or barium enema, or surgery.

Growth disorders

Take every opportunity to weigh and measure every child. Plot height and weight (and head circumference if concerns about growth or development) on centile charts. Correct the age of the child for prematurity at birth until age 2y if born <32wk, and 1y if born at 32–36wk gestation.

Faltering growth ➜ p. 850 **Obesity** ➜ p. 152

Calculating expected height Small parents have small children and tall parents have tall children—always calculate expected height of the child before deciding the child has short stature or excessive height.

Expected height = (mother's height + father's height) ÷ 2
Then: add 6cm for a boy or subtract 6cm for a girl.

❗ 3% of 'normal' children fall under the 3rd and 3% above the 97th centile.

Short stature Height <3rd centile. Mainly healthy children (80%) but may indicate physical or emotional problems—especially if both parents have heights >3rd centile or serial measurements show growth has fallen below that expected from the centile chart. *Causes*:

- Genetic Achondroplastic dwarfism; familial short stature; Turner's syndrome; familial growth delay (children have delayed pubertal growth spurt but eventually reach normal height)
- Physical Low birthweight conditions; endocrine causes, e.g. growth hormone deficiency, hypopituitarism, hypothyroidism, DM; chronic illness, e.g. severe asthma, heart disease, chronic infection
- Non-organic Poor nutrition; eating disorders; neglect (uncommon cause)

Assessment

- Ask how the child eats and about problems with feeding the child—appetite, food fads, special diets, quantities/times, snacks, etc.
- Ask about other physical problems, e.g. breathlessness, diarrhoea
- Examine the child carefully from top to toe looking for any physical abnormalities or signs of developmental delay
- Observe child's interactions, consider neglect
- Measure parental heights—small parents may have small children

Management Treat reversible causes. Continue to measure height and weight regularly. Refer to paediatrics if no cause for short stature, or an abnormality requiring specialist care, is found, or if, despite treatment of a reversible cause, the child fails to grow along his/her growth curve.

Pituitary dwarfism ↓ function of the anterior pituitary gland, causing short stature or failure to thrive. Skeletal maturation, assessed by bone age, is usually >2y behind chronologic age. *Causes*:

- Idiopathic
- Genetic
- Midline defect, e.g. cleft palate
- Pituitary tumour, e.g. craniopharyngioma

Management Specialist management is essential. Treatment is with growth hormone ± cortisol, thyroid hormone, and/or gonadal sex steroids.

❗ Growth hormone treatment ↑ risk of slipped femoral epiphysis.

Excess height Most tall children (height >97th centile) are from tall families. Pathological causes (rare) include: pituitary adenoma (gigantism), thyrotoxicosis, precocious puberty, Marfan's syndrome, and homocystinuria. Refer if much taller than predicted height or if growth deviates from growth curve.

Head growth At birth, head circumference is 32–37cm (term infant); the anterior fontanelle measures 2.5 × 2.5cm, becoming smaller until closing at 6–18mo. Most head growth occurs in infancy. Consider referral if head circumference crosses ≥2 centiles or is significantly out of proportion to body size.

- Microcephaly 1 in 1000 births. Small head out of proportion with the size of the body. Associated with developmental delay. *Causes*: genetic, intrauterine infection (e.g. rubella, CMV), chromosome abnormality, fetal alcohol syndrome, hypoxia
- Macrocephaly Large head circumference. *Causes*:
  - Hydrocephalus (➔ p. 878)—suspect if head circumference deviates from normal curve or if there are signs of ↑ intracranial pressure—refer
  - Megalencephaly—usually benign/familial—rarely associated with developmental delay
- Asymmetrical skull *Causes*: postural effects, e.g. children who sleep on one side (reassure—usually resolves with time); craniosynostosis (premature fusion of skull sutures—if suspected refer for prompt neurosurgical opinion)

Disorders of puberty

Delayed puberty Affects ~2% of young people. No pubertal changes by 13y in girls or 14y in boys, or no progression of puberty over 2y. May be constitutional (>50% boys) or pathological (80% girls). In all cases, refer to a paediatrician for further assessment. Pathological causes:

- Failure of the ovaries/testes (1° or hypergonadotropic hypogonadism)
- Failure of stimulation of normal gonads to produce sex hormones (2° or hypogonadotropic hypogonadism)

Precocious puberty Puberty before the normal age for the population (in the UK: <8y for girls; <9y for boys). Affects 4–5% girls. ♀:♂ ≈5:1. In all cases, refer for further investigation. *Types*:

- Central or true precocious puberty (80%). Premature activation of the hypothalamic–pituitary–gonadal axis. No pathological cause is found in 50–60% ♂ and 90% ♀. There may be a FH
- Pseudo-precocious puberty (20%). ↑ level of sex hormones in the absence of excess FSH or LH. There is usually a pathological cause

Other problems of puberty

- Asymmetric breast growth in girls Reassure that usually evens out by the time of full maturation
- Gynaecomastia in boys ~50% during puberty. Small, firm lump under one/both nipples. Reassure. Usually resolves without treatment
- Primary amenorrhoea (without delayed puberty) ➔ p. 682
- Premature pubarche (or adrenarche) Early appearance of pubic ± axillary hair and body odour without other signs of precocious puberty. If no other abnormalities of androgen excess, reassure

Information and support for children and parents

Child Growth Foundation ☎ 020 8995 0257 🖱 www.childgrowthfoundation.org

Other childhood endocrine problems

Addison's disease ➲ p. 338

Childhood diabetes mellitus (DM)^{N} *Incidence*: 2.5 per 1000 children. *Prevalence*: 1 in 500 <19y-olds. *Peak age at diagnosis*: 10–14y. 96% have type 1 DM.

Presentation of DM Usually presents with short history of polydipsia, polyuria, and weight ↓. May present with excessive tiredness or ketoacidosis.

Diabetic ketoacidosis (DKA) in children Potentially fatal—children with DKA can deteriorate rapidly. Presents with nausea/vomiting, abdominal pain, hyperventilation, dehydration, and/or ↓ conscious level.

> ⚠ *Initial management of suspected DM or DKA* Check capillary blood glucose in ANY child who presents with possible symptoms of DM or DKA. If >11mmol/L, refer immediately for paediatric assessment.
>
> Children taking insulin for DM can develop DKA with normal glucose. If they present with symptoms/signs of DKA, check blood ketones if monitor available. If ↑ ketones or unable to test, refer immediately for paediatric review.

Ongoing management Multidisciplinary and specialist led, usually involving insulin treatment (for type 1 DM) or metformin (for type 2 DM), education, psychological support, and monitoring for complications.

Further information

NICE (2015, updated 2016) Diabetes (type 1 and type 2) in children and young people: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng18

Congenital goitre Enlarged thyroid gland present at birth ± hypo- or hyperthyroidism. Hypothyroid babies are treated with thyroxine; if there is tracheal compression or hyperthyroidism treatment is surgical.

Hypothyroidism

Neonatal (congenital) hypothyroidism ➲ p. 830

Juvenile (acquired) hypothyroidism Usually due to autoimmune thyroiditis (Hashimoto's thyroiditis). Often insidious onset with ↑ weight, constipation, dry or coarse hair, and sallow, cool, or mottled coarse skin. There may also be developmental delay, growth retardation, delayed skeletal maturation ± delayed puberty. TFTs confirm diagnosis. Refer to paediatrics for specialist management. Treatment is with thyroxine replacement.

Hyperthyroidism

Neonatal hyperthyroidism Rare but potentially life-threatening. Occurs in infants of mothers with current or prior Graves' disease due to passage of autoantibodies across the placenta. *Presentation*:

- Feeding problems
- Faltering growth
- Vomiting/diarrhoea
- Irritability
- Tachycardia ± ↑ BP
- Exophthalmos
- Goitre
- Frontal bossing
- Microcephaly

Refer for specialist management. Affected infants generally recover in <4mo. Long-term consequences include premature fusion of the cranial sutures (craniosynostosis) and developmental delay.

Juvenile hyperthyroidism Usually Graves' disease (➔ p. 335). TFTs confirm diagnosis. Refer for specialist management. Treatment is with anti-thyroid medication. Spontaneous resolution in <2y is the norm.

Congenital adrenal hyperplasia (CAH) Also known as *adrenogenital syndrome* or *adrenal virilism*. Autosomal recessive trait due to absence or deficiency of any of the enzymes needed for synthesis of cortisol. Each enzyme block causes a characteristic deficiency. 2 patterns:

- Androgens accumulate causing virilization of an affected female fetus
- Androgen synthesis is impaired causing inadequate virilization of an affected male fetus (much rarer)

Presentation and management Ambiguity of the external genitalia. Less severe forms may go unnoticed until puberty. There may be a FH of CAH, ambiguous genitalia, or neonatal death. Rarely presents with Addisonian crisis (➔ p. 1083). Refer for specialist management. Treatment is usually with glucocorticoid ± mineralocorticoid replacement.

Male hypogonadism ↓ function of the testes. 3 types:

- Primary Damage to the Leydig cells impairs androgen (testosterone) secretion, e.g. Klinefelter's syndrome, anorchia (absent testes)
- Secondary (hypogonadotropic) Disorders of the hypothalamus or pituitary (e.g. panhypopituitarism) impair gonadotrophin secretion which may result in impotency and/or infertility
- Resistance to androgen action Presentation depends on age of onset:
    - *In utero*—ambiguity of genitalia or female appearance, small penis, incomplete testicular descent
    - *In childhood*—delayed or impaired puberty, impaired development of male sexual characteristics ± gynaecomastia
    - *In adulthood*—↓ libido, impotence, loss of muscle power, testicular atrophy, fine wrinkling of the skin around eyes and lips, sparse body hair, osteopenia, gynaecomastia

Management Refer for specialist investigation and treatment.

Androgen insensitivity syndrome (AIS) X-linked genetic disorder affecting 1 in 62,000 ♂ births. Abnormalities within androgen receptors result in the individual being genotypically male (46XY) but phenotypically female. External genitalia are female in complete AIS (CAIS), but are often ambiguous in partial AIS (PAIS). The testes fail to descend and are usually found in the groin—rarely within the abdomen. At puberty, breast development occurs and female contours form, but there is little or no pubic or axillary hair. Patients often present with primary amenorrhoea.

Management As diagnosis is often made in teenage years, young people with CAIS have usually been treated as female since birth. Specialist support is always required. Testes are usually removed as they have malignant potential, and oestrogens given to complete secondary sexual development.

⚠ Sexual ambiguity at the neonatal or 6-week check

- Be honest—do not guess the gender of the child
- Explain that there are rare conditions where girls may be virilized or boys undermasculinized, causing girls to look like boys and vice versa
- Arrange paediatric assessment as soon as possible for further investigations, gender assignment, and ongoing management

Funny turns and febrile convulsions

Funny turns Small children are often brought to the GP by their parents because they have had a funny turn. The major questions are: *Was the episode a fit? If so, what caused it? If not, then is there another serious underlying cause, e.g. heart disease?*

History A good history from a witness is essential. Ask:

- When did the attack happen and where?
- Were there any precipitating events or warning signs, e.g. viral illness; fever; strong emotions (was the child angry or upset?); head injury?
- What happened? Did the child lose consciousness or jerk limbs? If so, was the jerking generalized or restricted to one area of the body? What did the child look like during the attack, e.g. colour, floppiness? Did anything else happen during the attack, e.g. tongue biting?
- How long did the attack last?
- What happened after the attack? Was the child conscious straight away? Was there disorientation or drowsiness?

Also check

- General history—is the child well? Does the child have any ongoing medical problems? Past medical history—serious illness, neurological and/or developmental problems, heart problems
- Birth history—problems in pregnancy, birth trauma
- FH—epilepsy

Examination Full general/neurological examination; developmental milestones. Plot head circumference and weight on a centile chart.

Differential diagnosis

- Epileptic attack—febrile convulsion or childhood epilepsy (➔ p. 876)
- Non-epileptic attack

Non-epileptic attacks Usually self-limiting and harmless. Frightening for parents/carers, so education about likely duration and cause of attacks, and reassurance the child will come to no harm are important.

Simple blue breath-holding attacks Onset usually >6mo of age. Common. Provoked by frustration or upset. *Signs*: +ve Valsalva manoeuvre, cyanosis, stiffening, and loss of consciousness. No treatment needed—spontaneous recovery. Most children 'grow out' of the attacks by 3y.

White reflex asystolic (anoxic) attacks Most common from 6mo—2y. Usually triggered by minor injury or anxiety. *Signs*: vagal asystole, pallor, rapid loss of consciousness, stiffening, upward eye movement ± urinary incontinence. No treatment needed—spontaneous recovery.

Reflex syncope or vasovagal attacks ('faints') ➔ p. 525

Other causes are rare in children but include

- Cardiac arrhythmia—if recurrent loss of consciousness or collapse on exertion, refer for paediatric cardiology assessment
- Hyperventilation
- Benign monoclonus of infancy
- Benign paroxysmal vertigo
- Sleep phenomena
- Hypoglycaemia
- Fabricated or induced illness

Febrile convulsions Epileptic seizures provoked by fever in otherwise normal children. *Prevalence*: 3–5% of children aged 6mo–5y (peak age 18mo). There is often a FH. Seizures are usually brief (<5min) and generalized. *Causes (in order of ↓ frequency)*: viral infections; otitis media; tonsillitis; UTI; gastroenteritis; LRTI; meningitis; post-immunization.

Management of the fitting child ➔ p. 1080

Further management Children do not all require admission. Check temperature, assess respiratory/heart rate, capillary return, and level of consciousness. Examine for a source of infection. If there is no obvious cause of infection and not being admitted, dipstick urine to exclude UTI.

> ⚠ Complex convulsions are more likely to be provoked by a serious condition. Suspect serious pathology if a child has:
> - A prolonged (>15min) or focal febrile convulsion, or
> - Not recovered fully within <1h of a febrile convulsion

Arrange immediate paediatric assessment if

- First febrile seizure
- Complex seizure, the child was drowsy before the seizure, is irritable, systemically unwell or 'toxic', and/or the cause of the fever is unclear
- Symptoms/signs of meningitis (➔ p. 1056); petechial rash; recent or current treatment with antibiotics (may mask symptoms/signs of meningitis); or aged <18mo (meningitis may have non-specific signs)
- The cause of the fever requires hospital management in its own right
- Early review by a doctor is not possible, inadequate home circumstances, or the carer is anxious or unable to cope

For children not being admitted Reassure parents/carers that febrile convulsions do not harm the child. Antipyretics (e.g. paracetamol) do not prevent convulsions but are useful for symptom control. Advise parents to seek urgent medical help if the child deteriorates in any way, develops a non-blanching rash, fits again, or they are worried. Arrange early review (in <24h). Recommend that immunization schedules are completed.

Consider outpatient referral if

- Diagnosis of febrile convulsion is in doubt
- Febrile convulsions have been frequent, severe, and/or complex, and prophylactic treatment might be indicated
- The child is at ↑ risk of epilepsy, e.g. coexistent neurological or developmental conditions; history of epilepsy in a first-degree relative
- Parents/carers are anxious despite reassurance or request referral

Prognosis Febrile convulsions recur in subsequent febrile illness in ~30% of children. 1% of children having a febrile convulsion go on to develop epilepsy (compared to 0.4% of children who do not).

Further information

NICE (2012, updated 2018) The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care. 🖱 www.nice.org.uk/guidance/cg137

Childhood epilepsy

Childhood epilepsy is a susceptibility to continuing seizures. Prevalence ↑ with age from ~4 in 1000 children at 7y to ~5 in 1000 children at 16y. 60% of adult epilepsy starts in childhood.

Risk factors Neurological abnormalities or developmental delay; FH; past history of febrile convulsions—1% develop epilepsy.

Diagnosis Seizures, faints, and funny turns can be difficult to distinguish and diagnose (➔ p. 874). A reliable eye witness account is the key. If recurrent episodes ask parents to video on smart phone if possible.

⚠ Refer to a specialist paediatrician with training and expertise in epilepsy for diagnosis. All children who have had a first non-febrile seizure should be seen in <2wk. Consider referral to be seen in <48h if newly abnormal cerebellar or other central neurological function.

Management of the fitting patient ➔ p. 1080

Long-term management of epilepsy ➔ p. 548

Epileptic syndromes In children, epilepsy is considered in terms of the 'epileptic syndrome', characterized by a set pattern of seizure type(s) ± other features: physical appearance; age at onset; FH; associated learning disability and/or developmental delay; associated neurological findings; and EEG (should be undertaken in any child with a history of ≥2 epileptic seizures). Identifying a syndrome enables predictions about cause, severity, and prognosis.

❗ It is not possible to identify a syndrome in 30%—and symptoms/signs may take months to evolve until diagnosis can be made in others.

Benign epilepsy with centrotemporal spikes ('*Rolandic epilepsy*') 15–20% of childhood epilepsy. Starts aged 2–12y (peak age 7–10y)—usually stops by 13y. FH is common. Clonic, partial sensorimotor attacks affect the face, tongue, pharynx, hand, and arm. Most common on falling asleep (>½ have seizures only during sleep) or soon after waking. Secondary generalization to tonic–clonic seizures may occur. EEG is characteristic. Use of drug treatment depends on frequency and severity of seizures.

Juvenile myoclonic epilepsy (JME) 4–12% of childhood epilepsy. Age of onset 5–24y (peak age 10–16y). 50% have FH of epilepsy. Presents with sudden, brief, bilaterally symmetrical, and synchronous involuntary muscle contractions. Upper body > lower body. May cause the patient to throw objects or fall. Consciousness is often maintained. Frequently occurs soon after waking. Triggers may include light (50%), tiredness, or emotion. Generalized tonic–clonic seizures—often starting with a series of myoclonic jerks—appear <4y after onset of myoclonic seizures in ~90%. Absence seizures also occur in 15–30%. EEG is characteristic.

Management and prognosis Usually treated with sodium valproate or levetiracetam—fits may not be well controlled with medication. JME does not remit spontaneously Lifelong medication is needed—relapse rate is ~90% on withdrawal of antiepileptic medication.

⚠ Avoid prescribing sodium valproate to girls of childbearing potential. If essential, ensure a pregnancy prevention plan is in place and that information is provided about potential teratogenic effects both verbally and in writing.

Absence seizures ('*petit mal* epilepsy') 10–12% of childhood epilepsy. Age of onset 4–10y (peak age 5–7y); ♀ > ♂; ~15% have a FH. The child stops and may stare into middle space for a period of 4–20s. Can occur 50–100×/d. Deterioration in school performance may be the first sign. Separating absence attacks from daydreaming can be difficult. EEG is characteristic.

Management and prognosis 70% become seizure free with medication. ~15% go on to develop JME. ~10% have seizures in adult life.

Localization-related epilepsies Up to 30% of childhood epilepsy. Focal (partial) seizures that may be symptomatic (known underlying cause) or cryptogenic (cause not found). Clinical features, disabilities, and prognosis depend on cause and location of the brain abnormality.

Infantile spasms ('*West's syndrome*') Starts aged <1y (peak age: 4mo). Runs of tonic spasms—usually flexion ('salaam') spasms—occur every 5–10s. Characteristic EEG. Associated with loss of vision and social interaction. Treatment is with steroids and antiepileptics (usually vigabatrin). Poor prognosis—cerebral palsy (30–50%); cognitive disability (85%); 20% death rate.

Sturge–Weber syndrome Unilateral capillary naevus (port wine stain)—usually over the forehead/eyelid; epilepsy (90%); developmental delay (50%); hemiparesis and/or homonymous hemianopia (30%) and glaucoma in the affected eye. Treatment is supportive.

Lennox–Gastaut syndrome Severe, early-onset epilepsy (starts aged 2–6y) with multiple seizure types and typical EEG. Often treatment resistant.

Particular problems in children with epilepsy

- Developmental problems 25% have special educational needs; ~20% have learning disability. Specific cognitive disability (e.g. with reading or arithmetic) may have a serious impact on a child's education if not recognized
- Social stigmatization (perceived or experienced) Common. Children may have problems making friends because they cannot do everything other children do or have funny turns and are considered 'odd'. Other problems include: ↓ confidence, poor self-esteem, behaviour problems (e.g. conduct disorder, school refusal), dependence on others, anxiety, and depression
- Adverse effects of antiepileptic drugs For girls, discuss the risk to the unborn child (➔ p. 807) and interactions with contraception (➔ p. 728)
- Physical trauma May occur as a result of having a seizure

Further information

MHRA (2018, updated 2019) Valproate use by women and girls. 🖱 www.gov.uk/guidance/valproate-use-by-women-and-girls

NICE (2012, updated 2018) The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care 🖱 www.nice.org.uk/guidance/cg137

Information and support for children and parents

Epilepsy Action ☎ 0808 800 5050 🖱 www.epilepsy.org.uk

Hydrocephalus and neural tube defects

Hydrocephalus Accumulation of CSF and subsequent dilation of the cerebral ventricles as a result of ↑ production, ↓ absorption, or obstruction of CSF flow. Raised pressure can result in secondary brain damage.

Congenital hydrocephalus Present from birth. Prevalence ~1 in 1000 live births. May be a FH. Associated with ↑BP/pre-eclampsia in pregnancy, and neural tube defects. Specific causes include:

- Bickers–Adams syndrome (stenosis of the aqueduct of Sylvius)—associated with severe learning difficulties ± thumb deformity
- Dandy–Walker malformation
- Arnold–Chiari malformation type 1 and 2
- Congenital toxoplasmosis

Acquired hydrocephalus Occurs after birth. May be idiopathic. Specific causes include:

- Tumour, e.g. medulloblastoma, astrocytoma
- Intracranial haemorrhage, e.g. as a result of prematurity, following head injury, or after rupture of a vascular malformation
- Infection, e.g. meningitis
- ↑ venous sinus pressure, e.g. due to venous thrombosis, achondroplasia
- Excess vitamin A

Presentation in infants

- Faltering growth
- Developmental delay
- ↑ limb tone
- Irritability
- Vomiting
- Macrocephaly (>98th centile)
- Rapid ↑ in head circumference
- Separation of skull sutures; tense fontanelles
- Distended scalp veins
- ↓ conscious level
- Setting-sun sign (upper lids of both eyes are retracted and the white sclerae are visible above the iris)

Presentation in children

- **Acute onset** Headache, vomiting, papilloedema, impaired upward gaze
- **Gradual onset** Unsteady gait, large head (even if sutures are closed), 6th nerve palsy

Management If suspected, refer to paediatrics to be seen in <48h[N]. Diagnosis is made following CT scan (rarely USS through the anterior fontanelle in infants). Treatment is usually surgical by insertion of a shunt. A catheter is placed in the cerebral ventricle which drains CSF through a small reservoir just under the scalp, to either the right atrium of the heart (ventriculoatrial—VA shunt) or the peritoneal cavity (ventriculoperitoneal—VP shunt). Common complications of shunts include infection and shunt obstruction—if suspected, refer back to the specialist team managing the patient. ❗ All patients with a CSF shunt should have pneumococcal vaccination.

'Arrested' hydrocephalus Occurs when ICP returns to normal. The ventricles may remain dilated. Normal development resumes, but any damage caused before treatment remains.

Prognosis Untreated, hydrocephalus can be fatal. If treatment precedes irreversible brain damage, prognosis is good.

Neural tube defects Most neural tube defects are detected by routine antenatal USS. Inheritance is polygenic. Management is supportive. Types of defect:

Anencephaly Absent cerebral cortex and skull vault. Incompatible with life—those infants born alive, die within hours of birth.

Cranium defects Vary in severity from meningocele (meninges protrude through the defect) to encephalocele (brain tissue protrudes through the skull). Degree of long-term impairment depends on the size and location of the lesion.

Spina bifida The vertebral arch is incomplete. May be:

- **Open** (cystica) Herniation of the meninges (meningocele) ± spinal cord (myelomeningocele). Myelomeningocele is more common than meningocele and results in greater neurological deficit. Generally, the lower down the spine and the smaller the lesion, the less impairment there will be. Bladder and bowel problems are almost universal. Associated with hydrocephalus in the majority—especially after surgery
- **Closed** The defect is covered with skin/fascia. If just a defect in the vertebra (occulta) may have no consequences or result in neurological deficits, although these are usually mild. Sometimes caused by lipoma formation within the spinal canal during development which prevents vertebral closure (lipomyelomeningocele—15%). Not always detected on antenatal USS. Skin markers may be present, e.g. sacral pit, fatty lump, or overlying naevus or tuft of hair

Treatment Specialist treatment is always required and may involve surgery as well as support to manage neurological disabilities.

Associated conditions

- **Diastematomyelia** The spinal canal is bisected by a piece of bone or fibrous tissue. Treated surgically if causing symptoms
- **Tethered cord syndrome** As a child grows, the end of the spinal cord moves up within the spinal canal, to rest at ~L1 level by adulthood. If the spinal cord becomes tethered lower down, back pain and neurological symptoms affecting bowel/bladder function and legs may develop, particularly if rapid growth, e.g. at ~2y, aged 7–8y and around puberty. Treatment is surgical but later recurrences are common
- **Hydrocephalus** Commonly associated with spina bifida

❗ *Folate supplementation* ↓ risk of neural tube defect (open spina bifida, anencephaly, encephalocele) by 72%. For most women, recommend 0.4mg (400 micrograms) daily from when pregnancy is being planned until 13wk gestation. Recommend 5mg daily if:

- Previous child had neural tube defect
- Maternal/paternal history or other family history of neural tube defect
- Mother has coeliac disease, DM, BMI >30kg/m^2, or is taking anticonvulsants

Information and support for parents and children

Shine (spina bifida and hydrocephalus) 🖱 www.shinecharity.org.uk

Arthritis in children

Joint and limb pains are common in children. Arthritis is rare.

Presentation of arthritis in children

Older children Usually present with well-localized joint pains ± hot, tender, swollen joints.

Babies and young children May present with immobility of a joint or a limp, but the diagnosis can be extremely difficult.

Differential diagnosis of joint pains in children

- Juvenile idiopathic arthritis (JIA—incidence 1 in 10,000)
- Infections, e.g. TB, rubella, Lyme disease
- Rheumatic fever
- Henoch–Schönlein purpura
- Traumatic arthritis
- Hypermobility syndrome
- Leukaemia, lymphoma, or bone malignancy
- Sickle cell disease
- SLE and connective tissue disorders
- Transient synovitis of the hip (irritable hip)
- Septic arthritis
- Perthes' disease
- Slipped femoral epiphysis

Septic arthritis ➔ p. 494

Types of childhood arthritis Table 23.12

Management of children with arthritis

- If suspected, refer urgently to paediatrics for confirmation of diagnosis
- Once confirmed, ensure the child is referred to a specialist paediatric rheumatology unit to avoid long-term disability. These units have multidisciplinary facilities for rehabilitation, education, and surgical intervention (if necessary) and support both the family and the child
- NSAIDs and paracetamol help pain and stiffness, but corticosteroids and immunosuppressants (e.g. methotrexate) are often required for systemic disease. Monoclonal antibody therapy may be used if first-line disease-modifying agents are ineffective
- Ensure families apply for any benefits that might be available to them
- Tell families about self-help and support groups
- Support families in any applications made to adapt the home or school environment for the child's condition

Information and support for parents and children

Arthritis Research UK ☎ 0800 5200 520 🖱 www.arthritisresearchuk.org
Children's Chronic Arthritis Association ☎ 01905 745 595 🖱 www.ccaa.org.uk

Table 23.12 Childhood arthritis

Type of arthritis	Features
Oligoarthritis or pauciarticular-onset arthritis	
Persistent:	Most common form of JIA (50–60%) but still rare Peak age: 3y. ♀ >> ♂ Affects ≤4 joints—especially wrists, knees, and ankles. Often asymmetrical Associated with uveitis (often with +ve anti-nuclear antibody) which requires regular screening by slit lamp examination—rarely causes blindness Generally prognosis is good, with remission in 4–5y
Extended:	Chronic arthritis with an oligoarticular onset of the disease, which progresses to involve >4 joints. Joints tend to be stiff rather than hot and swollen
Still's disease (systemic onset JIA)	10% of JIA—only 1 in 3 have arthritis at the onset of the disease Aged <5y, ♀ = ♂ ; >5y ♀ > ♂ *Presentation*: ● Fever—high, swinging, early evening temperature for >2wk ● Rash—pink maculopapular rash with fever ● Musculoskeletal pain—arthralgia, arthritis, myalgia ● Generalized lymphadenopathy ● Hepatosplenomegaly ● Pericarditis ± pleurisy (uncommon) *Investigations*: blood—↑ ESR/CRP; FBC—↑ neutrophils, ↑ platelets. Autoantibodies are –ve
⚠ *Differential diagnosis*: malignancy—particularly leukaemia or neuroblastoma; infection	
Polyarticular-onset JIA	Starts with or without a preceding systemic illness at any age >1y, but most commonly in teenagers producing widespread joint destruction Symmetrical arthritis of hands, wrists, PIP joints ± DIP joints Rheumatoid factor is usually –ve (+ve in 3%—often teenage girls)
Enthesis-related JIA	Affects teenage and younger boys, often producing an asymmetrical arthritis of lower limb joints and tendon insertions. Heel pain and soft tissue swelling are common Associated with HLA-B27 and acute anterior uveitis Represents the childhood equivalent of adult ankylosing spondylitis. ~60% of childhood sufferers develop ankylosing spondylitis later in life.
Juvenile psoriatic arthritis	Polyarthritis affecting large and small joints, including fingers and toes. The arthritis can be very erosive Psoriasis may be present in the child or a first-degree relative (➲ p. 590)

Paediatric dermatology

Birthmarks

Strawberry naevus (capillary haemangioma)

- Not usually present at birth
- Occurs anywhere on the skin surface
- Starts as a small, red patch, then grows rapidly over a few months into a bright red vascular lump (Figure 23.5)
- After initial growth, the naevus stays the same size for 6–12mo, then involutes and disappears, usually by 5–7y
- No treatment is needed. Parents may need considerable reassurance
- If interfering with feeding, breathing, or vision or if ulcerating—refer for treatment with oral or topical beta blockers, intralesional steroids, or laser

Port wine stain (naevus flammeus)

- Present at birth
- Irregular red/purple macule which often affects one side of the face (Figure 23.5)
- Permanent—may become darker and lumpy in middle age
- May be associated with other abnormalities, e.g. intracranial vascular malformation (Sturge–Weber syndrome—➔ p. 877)

Salmon patch (stork mark)

- The most common vascular naevus (~50% neonates)
- Small, telangiectatic lesion forming a pink macule—most commonly at the nape of the neck or on the upper face (Figure 23.5)
- Facial lesions resolve spontaneously—those on the neck may persist. No treatment is needed

Mongolian blue spot

- Bluish discoloration of the skin (Figure 23.5); usually over buttocks and lower back in dark-skinned babies
- Of no clinical significance but may occasionally be mistaken for bruising and non-accidental injury
- Usually disappears by 1y

Congenital melanocytic naevi ~1.5% of neonates.

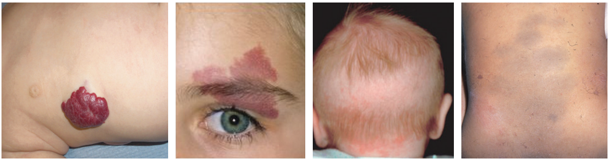
- Noted at birth as raised nodules or plaques of black or brown
- May be hairy, irregular, and single or multiple
- Classified by size: <1.5cm—small; 1.5–20cm—medium; >20cm—large
- There is a risk of malignant change to melanoma—the larger the naevus the greater the risk
- Laser therapy can improve cosmetic appearance

Nappy rash Most common type of nappy eruption. Usually seen in young infants—rare >12mo. An irritant dermatitis due to skin contact with urine or faeces.

Presentation Glazed erythema in the napkin area, sparing skinfolds. Secondary bacterial or fungal infection is common.

Differential diagnosis

- Seborrhoeic eczema
- Candidiasis
- Napkin psoriasis



Strawberry naevus *Port wine stain* *Salmon patch* *Mongolian blue spot*

Figure 23.5 Birthmarks

(a) Reproduced from Burge S. *et al*., Oxford Handbook of Medical Dermatology, second edition (2016) with permission from Oxford University Press; (b) Reproduced from Tasker R.C. *et al*., Oxford Handbook of Paediatrics, second edition (2013) with permission from Oxford University Press; (c) Reproduced from Mulliken J.B. *et al*., Mulliken and Young's Vascular Anomalies: Hemangiomas and Malformations (2013) with permission from Oxford University Press; (d) Reproduced from Lewis-Jones S., Paediatric Dermatology (2010) with permission from Oxford University Press.

Management Advise parents to keep the nappy area dry and to give baby as much time as possible with the nappy off. Apply moisturizer as soap substitute. Apply a barrier cream between nappy changes—although this may interfere with the action of some modern nappies. Topical treatment with an antifungal combined with hydrocortisone is effective if the nappy rash is not clearing.

Infantile seborrhoeic eczema Starts in the first few weeks of life. Affects body folds—axilla, groins, behind ears, neck ± face/scalp ('*cradle cap*'). Flexural lesions present as moist, shiny, well-demarcated scaly erythema. On the scalp, neck, and behind the ears, a yellowish crust is usual. Treat flexural lesions with emollients and 1% hydrocortisone ointment, or with clotrimazole and hydrocortisone cream. Scalp lesions respond to OTC cradle cap creams.

Candidiasis Frequently complicates nappy rash or infantile seborrhoeic eczema. Erythema, scaling, and pustules involve the flexures. There may be associated satellite lesions. Treatment is with a topical antifungal, e.g. clotrimazole.

Juvenile plantar dermatosis Presents with red, dry, fissured, and glazed skin principally over the forefeet. Sometimes involves the whole sole. Usually starts in primary school years and resolves spontaneously in mid-teens. Due to wearing socks and/or shoes made from synthetic materials. Emollients help but topical steroids are ineffective. Advise cotton socks and leather shoes.

Accessory nipples Commonly seen on the milk line in both male and female infants. Usually small and inconspicuous. No treatment is needed.

⚠ **Staphylococcal scalded skin syndrome** Rare but serious skin infection seen in infants/young children. Caused by staphylococcal infection releasing epidermolytic toxin causing widespread erythema/blistering. The epidermis shears off giving the appearance of scalded skin. Admit as a paediatric emergency for IV antibiotics and supportive treatment. ❗ Can also affect immunosuppressed adults.

Urticaria ➔ p. 586 **Acne** ➔ p. 588 **Scabies** ➔ p. 613
Psoriasis ➔ p. 590 **Warts** ➔ p. 608 **Head lice** ➔ p. 612
Atopic eczema ➔ p. 578 **Molluscum contagiosum** ➔ p. 609

Further information

Primary Care Dermatology Society Clinical A-Z list. 🖱 www.pcds.org.uk/a-z-clinical-guidance/clinical-a-z-list

Childhood cancer

Every year in the UK, 1800 children are diagnosed with cancer; 260 children/y die from cancer. Acute leukaemia is most common (1 in 3 cases) followed by brain tumour (1 in 4). Diagnosis of childhood malignancy is challenging as GPs rarely see children with cancer, so childhood cancers are unfamiliar to them. Always have a high index of suspicion; if in doubt refer for a specialist opinion.

Referral for suspected childhood cancer Box 23.1

Specific childhood cancers covered elsewhere

- Acute leukaemia ➔ p. 652
- Lymphoma ➔ p. 656
- Brain tumour ➔ p. 532
- Sarcoma ➔ p. 479
- Gonadal tumours—testicular (➔ p. 442); ovarian (➔ p. 698)

Neuroblastoma Tumour derived from neural crest tissue. 90 cases/y in the UK—6% of all paediatric tumours. Neuroblastoma tends to affect children aged <4y (50% <2y; 90% <9y). *Sites*: adrenal medulla—50%; abdominal sympathetic ganglia—25%; chest—20%; pelvis—5%; neck—5%. Presentation is variable and non-specific—depends on site of the tumour and extent of metastases. ~½ present with metastatic disease:

- **General effects** Pallor, fever, anorexia and weight ↓, faltering growth, diarrhoea, irritability, flushing, ataxia
- **Local effects** Abdominal mass (or thoracic mass on CXR); local spread may cause paraplegia or cauda equina syndrome. Infants <6mo may have rapidly progressive intra-abdominal disease
- **Metastases** Lymphatic and haematogenous spread, particularly to liver, lungs, and bone, is common. *Associated symptoms*:
  - Bone pain ± pathological fracture
  - Breathlessness
  - Periorbital bruising ('black eye'), proptosis, or Horner's syndrome
  - Firm skin nodules (usually babies—'blueberry muffin appearance')

Treatment is with surgery, chemotherapy, and/or radiotherapy. Early-stage disease: 95% 5y-survival; late-stage disease: 20% 5y-survival. Children aged <1y at diagnosis or with extra-abdominal tumours have better prognosis.

Wilms' nephroblastoma 80 cases/y in UK. Kidney tumour composed of primitive renal tissue; L > R; bilateral in 10%. Usually affects children <5y old (peak age: 2–3y). ♂ > ♀. 10% occur in children with rare syndromes, e.g. Beckwith–Wiedemann syndrome, aniridia, or hemihypertrophy. A few cases are familial. May present with metastases:

- **General effects** Fever, anorexia and weight ↓, anaemia
- **Local effects** Unilateral abdominal mass ± pain ± unexplained haematuria
- **Metastases** 10% at presentation—liver, lungs, or bone (rare)

Treatment is with surgery, chemo- and/or radiotherapy. 5y survival is 90% (higher for early-stage tumours).

Retinoblastoma Rare eye tumour. 40 cases/y in UK. 40% are aged <1y at diagnosis; 95% are <5y. May be familial (10%—dominant inheritance) when the tumour is usually bilateral; genetic testing is available. May be detected as a white pupillary reflex seen at developmental screening, or present with squint or inflammation of the eye. Treatment options include thermotherapy, brachytherapy, chemotherapy, or surgery. 5y survival is 100%.

Box 23.1 Referral of children with suspected cancer[N]

❗ Some congenital syndromes are associated with ↑ cancer risk (e.g. Down's—leukaemia; neurofibromatosis and tuberous sclerosis—CNS tumours).

Admission/referral to be seen immediately Any child with unexplained hepatosplenomegaly or unexplained petechiae.

Investigations

Check FBC (within 48h) If

- Pallor
- Persistent fatigue
- Unexplained bruising/bleeding
- Generalized lymphadenopathy
- Unexplained/persistent fever or infection
- Persistent/unexplained bone pain (additionally, consider X-ray)

❗ If FBC/blood film shows leukaemia, admit or arrange for same-day specialist paediatric assessment.

Arrange X-ray (within 48h) If persistent/unexplained bone pain (additionally, consider FBC), or unexplained bone swelling.

Arrange USS (within 48h) If any unexplained lump that is ↑ in size.

Urgent paediatric referral (to be seen in <48h) If a child has:

- Unexplained lymphadenopathy or splenomegaly, particularly if associated with night sweats, weight ↓, pruritus, fever, or shortness of breath
- Abdominal mass/enlarged abdominal organ
- Newly abnormal cerebellar or other central neurological function
- Unexplained, visible haematuria
- X-ray or USS suggesting possibility of bone or soft tissue sarcoma

Urgent ophthalmology referral (to be seen in <2wk) If absent red reflex (leucocoria).

Consider paediatric referral Where there is persistent parental anxiety, even when a benign cause of a child's symptoms is likely.

Ongoing care Most children with cancer embark on intensive treatment regimens managed by tertiary oncology units sharing care with local hospitals. They have direct access to advice/admission via those units.

⚠ Any febrile episode in a neutropenic child requires immediate referral to a specialist unit. Chickenpox is particularly serious—seek specialist advice from the treating unit if the patient has contact with another child with chickenpox.

The GP's role ➜ p. 900. Keep in touch and up to date with progress. Provide support to the child/family and advice on benefits/services that might be useful. Ensure prompt supply of prescriptions.

Palliative care ➜ p. 904

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Information and support for children and families

Childhood Eye Cancer Trust (CHECT) 🖱 www.chect.org.uk
Children with Cancer UK 🖱 www.childrenwithcancer.org.uk
CLIC and Sargent ☎ 0300 330 0803 🖱 www.clicsargent.org.uk
Neuroblastoma Society ☎ 020 8940 4353 🖱 www.neuroblastoma.org.uk

Behaviour problems

GPs are commonly asked to 'sort out' behaviour problems of children by parents who feel they can no longer cope. 2–10% of all children have behaviour problems depending on how problems are defined and measured.

Differentiation between normal behaviour and behavioural problems can be difficult—especially if you do not know the child or family well. A significant problem is more likely:

- When the behaviour is frequent and chronic
- When >1 problem behaviour occurs, *and*
- If behaviour interferes with social and cognitive functioning

There is no right or wrong way to deal with these problems and the approach outlined in Figure 23.6 is just one way to tackle them.

Managing the problem For simple problems, parental education, reassurance, and a few specific suggestions tailored to the situation are often sufficient. Follow-up is important to ensure that the problem is resolving. If simple measures are not succeeding within 3–4mo, consider referral to other agencies, e.g. health visitor, school nurse, parent support programmes, child and adolescent mental health service (CAMHS). Specific behavioural techniques include:

Behaviour modification A learning process requiring caregivers to set consistent rules and limits. Parents should try to minimize anger when enforcing rules and ↑ +ve contact with the child.

Discipline Ineffective discipline may result in inappropriate behaviour. Scolding may briefly control a child's behaviour if used sparingly but may ↓ the child's sense of security and self-esteem. Physical punishment and threats to leave or send the child away are damaging. *Options*:

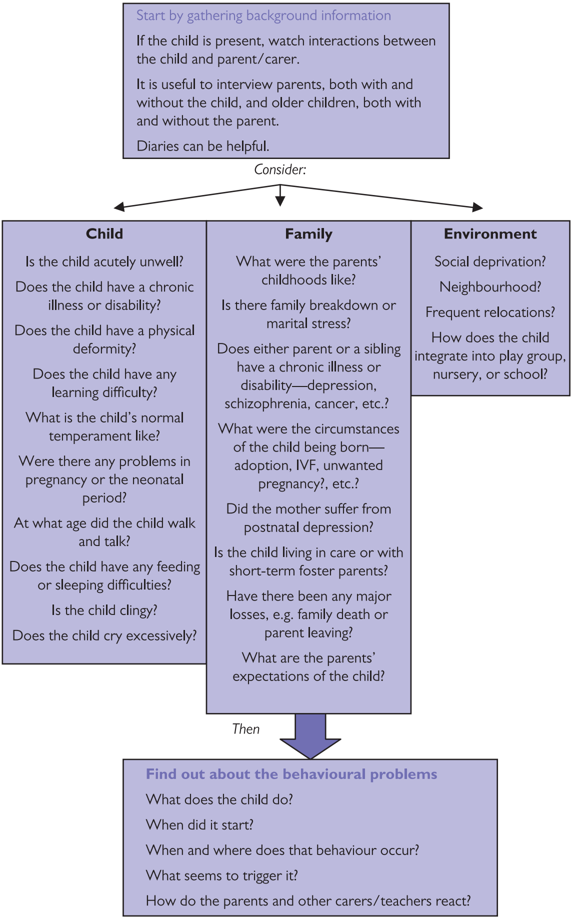
- **+ve reinforcement for appropriate behaviour** This is a powerful tool for controlling a child's behaviour with no adverse effects
- **Time-out procedure** The child must sit alone in a dull place for a brief period. Time-outs are a learning process for the child and are best used for controlling a single inappropriate behaviour or a few at one time

Breaking vicious circle patterns The child's behaviour (be it normal for that developmental stage or abnormal) evokes a response in the parent or carer which provokes the child to behave in that manner further—thus generating another response from the parent. Try to identify vicious circle patterns and suggest alternative parental responses which make the behaviour futile.

Sleep problems ➔ p. 890
Toilet training ➔ p. 892
Feeding problems ➔ p. 850
Hyperactivity ➔ p. 894
Childhood depression ➔ p. 899

Information and support for children

Childline 24h confidential counselling service. ☎ 0800 1111 🖱 www.childline.org



Start by gathering background information

If the child is present, watch interactions between the child and parent/carer.

It is useful to interview parents, both with and without the child, and older children, both with and without the parent.

Diaries can be helpful.

Consider:

Child

Is the child acutely unwell?

Does the child have a chronic illness or disability?

Does the child have a physical deformity?

Does the child have any learning difficulty?

What is the child's normal temperament like?

Were there any problems in pregnancy or the neonatal period?

At what age did the child walk and talk?

Does the child have any feeding or sleeping difficulties?

Is the child clingy?

Does the child cry excessively?

Family

What were the parents' childhoods like?

Is there family breakdown or marital stress?

Does either parent or a sibling have a chronic illness or disability—depression, schizophrenia, cancer, etc.?

What were the circumstances of the child being born—adoption, IVF, unwanted pregnancy?, etc.?

Did the mother suffer from postnatal depression?

Is the child living in care or with short-term foster parents?

Have there been any major losses, e.g. family death or parent leaving?

What are the parents' expectations of the child?

Environment

Social deprivation?

Neighbourhood?

Frequent relocations?

How does the child integrate into play group, nursery, or school?

Then

Find out about the behavioural problems

What does the child do?

When did it start?

When and where does that behaviour occur?

What seems to trigger it?

How do the parents and other carers/teachers react?

Figure 23.6 Assessment of childhood behaviour problems

Parent information and support

Green C (2006) *New Toddler Taming: A Parents Guide to the First Four Years*. London: Vermilion. ISBN: 9780091902582.

Parentline ☎ 0808 800 2222 🖱 www.familylives.org.uk

Phelan T (2016) *1-2-3 Magic: Effective Discipline for Children 2–12*. Naperville, IL: Sourcebooks. ISBN: 9781492629887.

Phelan T (2016) *1-2-3 Magic Teen: Communicate, Connect, and Guide your Teen to Adulthood*. Naperville, IL: Sourcebooks. ISBN: 9781492637899.

Excessive crying Babies vary considerably in the amount they cry and ease with which they are soothed. Likewise parents vary in their ability to tolerate a crying baby. Babies cry for many reasons—discomfort, hunger, loneliness, separation, boredom, etc. If a baby is crying excessively:

- Take a history from the parent—when does the baby cry, can he/she be consoled, what do the parents do when the baby cries?
- Examine fully from head to toe to exclude causes of discomfort, e.g. nappy rash, otitis media, eczema
- Check the baby is growing along his/her centile line
- Consider family stress (including postnatal depression—➔ p. 819) as a reason why the parent cannot tolerate the crying
- Treat any underlying cause found and support the family. Information about behavioural techniques is available from Cry-sis

Parent information and support

Cry-sis Support for families with crying and sleepless babies. ☎ 08451 228 669 🖱 www.cry-sis.org.uk

Rhythmic behaviour Head rocking or banging, thumb sucking, self-stimulation, baby behaviour, and many other variants all occur during normal development. They usually appear if the child is tired, uncertain, or anxious. Reassure parents. Most resolve spontaneously.

Fears and phobias

Fears

- Of the dark, monsters, and spiders are common in 3–4y-olds
- Of injury and death are more common in older children
- Statements made by the parents in anger or jest may be taken literally by preschool children and can be disturbing
- Frightening stories, films, or television may be upsetting and intensify fears

Phobias Cause persistent, unrealistic, yet intense anxiety in reaction to external situations or stimuli.

Management Normal developmental stage-related fears must be differentiated from true phobias. If the phobia is intense and interferes with the child's activity, or if the child does not respond to simple reassurance, refer to CAMHS.

School refusal and truancy

School refusal Children may refuse to go to school showing signs of distress or anxiety, or recurrently complain of abdominal pain, nausea, or other symptoms. Parents are aware of absence and child stays at home. Often school refusal is a form of separation anxiety, although it may be due to a problem at school, e.g. problems interacting with the teacher or friends, bullying, or learning difficulties. Advise parents to consult the school and agree a consistent approach, which may involve reward charts, and peer, pastoral, and/or parental support. CAMHS input may be needed if anxiety/depression is a prominent feature. Relapses can occur if the child is absent or after holidays. The school may be able to access support through educational welfare officers, parent support advisors, counsellors, or the school nurse.

Truancy Often older children. There is no anxiety. The child does not usually remain at home and absence is often concealed from parents. Often associated with antisocial behaviour. Speak to parents and child together and separately. Try to ascertain if there is a genuine reason why the child avoids school. Treat or refer on for any physical or mental health condition identified. Liaise with the school as for school refusal.

Conduct disorders Poor behaviour (e.g. aggression, destructive tendencies, and antisocial behaviour) are common complaints. Tolerance varies from family to family. Try simple strategies such as rewarding good behaviour and ignoring poor behaviour ± 'time-out' strategies (younger children—➔ p. 886). If not succeeding, consider referral to CAMHS.

Tics Sudden, rapid, repetitive non-rhythmic motor movements of no apparent purpose. Commonly involve facial grimacing, head movements, or shoulder movements. Average age of onset is ~5y. Tics are present at some point in ~4% of children. Often a FH is present. Tics come and go and vary in intensity and frequency. They are often precipitated by stress. Most disappear during adolescence but in 20% they persist into adulthood.

Gilles de la Tourette syndrome ♂:♀ ≈4:1. Characterized by multiple motor and vocal tics—sometimes obscene. There may also be repetitive blinking, nodding, gesturing, echoing of speech, and/or stuttering. Begins in childhood. Associated with obsessive–compulsive disorder (OCD) and attention deficit hyperactivity disorder (ADHD). Probable genetic aetiology.

Management Refer to CAMHS for specialist treatment and support. Frequency and severity of tics varies from person to person and throughout life. Many affected children/young people improve after puberty. Treatment is with psychoeducation, behavioural techniques, and, in severe cases, clonidine or antipsychotic medication may help (usually specialist initiated). Treat any associated OCD (➔ p. 974) or ADHD (➔ p. 895).

⚠ **Behavioural problems and maltreatment**[N] Consider maltreatment (➔ p. 902) if there is behavioural change inconsistent with age/development that is not explained by a medical condition (e.g. ADHD, autism) or stressful situation (e.g. parental separation). In particular, suspect maltreatment if:

- A child repeatedly scavenges, steals, hoards, or hides food, or
- There is precocious, coercive, or unusual sexualized behaviour

Refugee children Many child refugees have traumatic backgrounds. Approach children with sensitivity and consider involving specialist CAMHS and refugee support services early. Recognize that children may be trafficked and exploited; refer if suspected to social services ± police.

Further information

NICE (2009, 2017) Child maltreatment: when to suspect maltreatment in under 18s. 🖱 www.nice.org.uk/guidance/cg89

Parent information and support

Parentline ☎ 0808 800 2222 🖱 www.familylives.org.uk

Information and support for children

Childline 24h confidential counselling service. ☎ 0800 1111 🖱 www.childline.org

Sleep problems in children

Sleeping patterns and habits of children vary considerably and should only be regarded as problems when they are presented as such by the family. First, take a careful history. *Ask about*:

- **Medical problems** e.g. night cough related to asthma, itching from eczema, obstructive sleep apnoea. Treat appropriately
- **Physical problems** e.g. hunger or cold
- **Night terrors**
- **Sleep pattern** Usually one of:
  - Difficulty settling
  - Waking during the night
  - Waking early in the morning
- **Amount of daytime sleep**

General advice In all cases it is helpful to recommend turning off electronic screens >1h before bedtime, a regular calming bedtime routine (e.g. bath, story, cuddle, bed), and minimal fuss when a child does wake at night, e.g. try to settle back to sleep without taking out of cot, not rewarding waking with games, snacks, etc.

Resistance to going to bed The baby/child who cries incessantly when put to bed is a common problem with a peak age of 1–2y. The child cries when left alone or climbs out of bed and seeks the parents. *Causes include*:

- Separation anxiety
- Increasing attempts by the child to control his/her environment
- Long naps late in the afternoon
- Rough, overstimulating play before bedtime
- A disturbed parent–child relationship and/or tension in the home

Behavioural management Not recommended for babies aged <6mo. Letting the child stay up or punishing the child are ineffective. *Options include*:

- **Leaving the child to cry** May work and the crying diminishes after a few nights—but very hard for parents to do and can be impossible if they are in shared accommodation
- **Controlled crying** The child is put to bed and the parent leaves the room. If the child cries, he/she is left for 2–10min before the parent returns to check with minimum fuss and then leaves. Length of time before returning is gradually ↑. Effective and easier for parents than leaving the child to cry
- **Gradual withdrawal** Staying with the child until he/she sleeps but gradually withdrawing proximity, e.g. sit on the bed with the child; after a few nights sit next to the bed, then nearer door, etc. until the child learns to go to sleep alone. This method is gentler but may take longer

Waking during the night Occurs in half of all children aged 6–12mo and may be related to separation anxiety. In older children, episodes often follow a stressful event (e.g. moving home, illness).

Management Allowing the child to sleep with the parents, playing with, feeding, or punishing the child usually prolongs the problem.

- Try the methods used for resistance to going to bed—but advise parents to always check to see that the child is not ill/needing a clean nappy, etc. before being left to cry
- Scheduled waking has been shown to be effective—the child is woken 15–60min before the time he/she usually wakes and then resettled
- If a child wakes early, another strategy is to make toys or books accessible. The child can then amuse him-/herself for a period of time without disturbing the parents. Advise fitting a stair gate across the child's bedroom door to prevent the child coming to any harm
- Use of sedatives, e.g. promethazine (children >2y), is often discouraged but can be useful as a short-term measure when parents feel desperate
- Children with neurodevelopmental disabilities often have disturbed sleep, melatonin may help (specialist-only prescription)

Nightmares Occur during rapid eye movement (REM) sleep. Nightmares can be caused by frightening experiences (e.g. scary stories, television violence), particularly in 3–4y-olds. The child usually becomes fully awake and can vividly recall the details of the nightmare. An occasional nightmare is normal, but persistent/frequent nightmares need evaluation by an expert.

Sleepwalking (somnambulism) Involves walking clumsily, usually avoiding objects. The child appears confused but not frightened. 15% of children aged 5–12y have sleepwalked one or more times. It is most common among school-aged boys and may be triggered by a stressful event.

- Advise parents/carers not to try to wake the child
- If the child is in danger, gently steer him/her away from any harm
- If the child sleepwalks frequently, consider action to prevent the child coming to harm while sleepwalking, e.g. stair gate across bedroom door
- If the sleepwalks occur repeatedly at the same time, waking the child ~15min before the predicted time can break the cycle

Night terrors Sudden awakening with inconsolable panic and screaming. Usually occurs in the first 1–3h of sleep. Episodes last seconds → minutes. *Features*:

- Blank or confused stares
- Incomplete arousal with poor responsiveness to people
- Amnesia for the episode

Night terrors are most common in children aged 3–8y and require no treatment apart from simple reassurance. Advise parents not to wake the child as this ↑ the disturbance. If frequent consider waking the child before episodes occur and keeping the child awake for a few minutes to break the cycle. If the terrors persist beyond 8y, consider a diagnosis of temporal lobe epilepsy.

Parent information and support

Cry-sis Support for families with crying and sleepless babies. ☎ 08451 228 669 🖱 www.cry-sis.org.uk

Parentline ☎ 0808 800 2222 🖱 www.familylives.org.uk

Toilet training

Most children can do without nappies by day from 2–3y and by night from 3–5y. How to approach toilet training will vary from child to child.

General rules

- **Wait until the child is ready** This usually means that the child can indicate to the parent that he/she is going to the toilet and has shown an interest in using the potty or toilet. It is helpful to have a potty or child's toilet seat to put on the normal toilet for the child to become familiar with before starting toilet training
- **Pick a good time** When the child can have a few days at home without nappies in an environment where accidents do not matter. Make sure the child has plenty of spare clothes available
- **Keep the potty handy or stay within easy reach of the toilet** If a child says he/she wishes to go, sit the child immediately on the toilet. Reward any result with praise. Do not punish the child for accidents
- **Play safe** Until the child (and parent) are confident in the child's ability to use the toilet, continue using nappies when out and at night. Take the child to the toilet at night before bedtime. When dry nappies are consistently noted in the mornings, try the child without nappies at night—a plastic sheet on the mattress is a good idea. Even when a child has been dry day and night for some time, accidents are common if the child is tired, unwell, or unsettled (whether excited or unhappy)

❗ If the child does not succeed within a few days, either try training pants or revert to nappies and try again at a later date.

Nocturnal enuresis[N] Involuntary voiding of urine during sleep on >2 nights/wk. Affects 8% of children aged 4½y and 1.5% at 9y. ♂ > ♀. Tends to run in families. Distressing for child and parents with effects on emotional and social well-being. Cause is not fully understood:

- 1–2% have an underlying physical abnormality—usually UTI, constipation, or DM (rarely congenital anomalies, diabetes insipidus, or pelvic mass). Exclude with history and examination—consider urinalysis if recent onset, daytime symptoms, or symptoms/signs of ill health
- Occasionally caused by emotional distress or recent illness—consider if the child has been dry for 6mo prior to onset. Explore possible triggers that may need treating. Consider maltreatment, particularly if parents blame/punish the child despite advice that it is not the child's fault

Management See Table 23.13. Often resolves with time.

Further information

NICE (2010) Bedwetting in under 19s. 🖱 www.nice.org.uk/guidance/cg111

Information for parents of children with enuresis

ERIC Enuresis resource and information. ☎ 0845 370 8008 🖱 www.eric.org.uk

Table 23.13 Management of enuresis

Method	Features
Advice and information	Reassure parents and children that enuresis is not the child's fault. In particular, advise: • That punitive measures should not be taken • On ways to limit the impact of bedwetting, e.g. bed protection • On regular toileting 4–7×/d and before bed • On appropriate fluid intake (1–1.4L/d in a child aged 4–8y) • To avoid caffeine • About self-help organizations/sources of support, e.g. ERIC
Reward systems	For example, star chart Use for agreed behaviour (e.g. going to the toilet before bed) rather than a dry night
Lifting and waking	Waking the child to go to the toilet several hours after going to bed Useful short term but will not promote long-term dryness. However, young people who have not responded to treatment may find self-waking with an alarm clock useful
Enuresis alarms Refer to the school nurse or paediatric enuresis clinic	An alarm is triggered to wake the child when urine makes contact with a sensor. With time the child wakes in response to bladder contractions rather than the alarm First-line treatment when advice/rewards fail. Usually some effect in <4wk but may take months until the child is completely dry at night. Continue until 2wk of dry nights—restart if relapses
Desmopressin Usually specialist initiated	Synthetic version of antidiuretic hormone. Taken at night Side effects include headache, nausea, nasal congestion, nose-bleed, sore throat, cough, flushing, and mild abdominal cramps ❗ There is a risk of water overload—advise only one mug of fluid from 1h before desmopressin dose to 8h afterwards Used when alarm is unsuccessful or unsuitable or in combination with an alarm. If effective continue for 3mo, then withdraw to assess if still needed
Anticholinergics/ imipramine	Specialist initiation only for children who have not responded to other treatments

Encopresis Most children are continent of faeces by 4y. Faecal soiling after this age usually occurs during the day. If:

- Soft stool oozes out, causing constant soiling, consider overflow incontinence secondary to constipation (➔ p. 866)
- A firm stool is passed occasionally in the toilet but usually in the pants, developmental delay (either mental or social) is likely. Try a firm, consistent training programme
- Bowel control, but the child passes stool in unacceptable places/smears faeces. The cause is usually emotional. Emotional/behavioural support is needed and, if not improving, consider child and adolescent mental health service referral. Consider maltreatment (➔ p. 902)

Poor progress at school

~20% of school-aged children require special educational services at some point in their schooling. ♂:♀ ≈5:1. *Consider*:

- Is a child's physical illness affecting school work (e.g. asthma, eczema)?
- Is medication affecting academic performance (e.g. anticonvulsants)?
- Is the family stable or is there family upset?
- Does another family member have a chronic/life-threatening illness?
- Is the child's home environment conducive to doing school work?
- Is this school refusal? (➔ p. 888)
- Is the child happy at school?
- Is there a problem with vision or hearing?
- Is the child of normal intelligence?
- Does the child interact socially with adults and other children?
- Have developmental milestones been met?
- Does the child have specific difficulty with certain aspects of his/her school work (e.g. mathematics, reading, writing)?

Learning disability ➔ p. 897

Specific learning disorders

Dyslexia and dyscalculia Difficulty in information learning/processing. Intelligence is often normal/high and the child appears bright and alert. There may be a FH.

- Dyslexia Difficulty with letters/words, resulting in problems with reading, writing, and spelling. Affects up to 10% of the population. ♂ > ♀
- Dyscalculia The core problem is difficulty handling numbers and mathematical concepts. Less common than dyslexia

There is considerable overlap between dyslexia, dyscalculia, and also dyspraxia. If suspected, liaise with the child's school. Formal testing by an educational psychologist can confirm the diagnosis. Specialized educational assistance and support are helpful.

Developmental coordination disorder (dyspraxia) Difficulty affecting movement/coordination. Affects 2–6% of the population; ♂ > ♀. Intelligence is often normal/high. Features are variable but include:

- Clumsiness
- Poor posture
- Awkward gait
- Reading and writing difficulties
- Difficulty holding a pen or pencil properly
- Poor short-term memory
- Poor body awareness
- Confusion about which hand to use
- Difficulties throwing/catching balls
- Poor sense of direction
- Difficulty hopping, skipping, and/or riding a bike
- Slow to learn to dress and feed

Diagnosis involves specialist community paediatric multidisciplinary assessment. Treatment is supportive.

Speech and language delay ➔ p. 837

Hyperactivity Difficult to define as claims a child is hyperactive often reflect the tolerance level of the person complaining. Active children with shorter-than-average attention spans create management problems.

Hyperactivity may have an underlying cause (e.g. an emotional disorder, CNS dysfunction, a genetic component) or may be an exaggeration of normal temperament. Often it is stage related—support until that stage has passed. Simple behaviour management techniques help (➔ p. 886).

Attention deficit hyperactivity disorder (ADHD)^{N}

- Common neurodevelopmental disorder interfering with normal social functioning, learning, and development
- Aetiology is multifactorial. Prematurity and substance misuse in pregnancy are risk factors; environmental and genetic factors contribute
- Affects up to 9% of the school age population in the UK; ♂:♀ ≈6:1
- Other neurodevelopmental, neurological, mental health, and learning difficulties may coexist
- Long-term ADHD is associated with low academic achievement, substance misuse, unemployment, and antisocial tendencies

Presentation ❗ Many of these behaviours are seen in normal children.

- Inattention Poor attention to detail and organization of tasks; appears not to listen; easily distracted; forgetful; lack of concentration
- Impulsivity Lack of social awareness; shouts out answers to questions; difficulty waiting (unable to take turns or wait in a queue); excessive talking—interrupts others; lack of social awareness
- Hyperactivity Fidgets; inappropriate running/climbing/leaving seat

Assess duration of symptoms, extent of functional impairment, and whether problems occur in different settings/areas of functioning—a school report can be useful. ❗ Diagnosis of ADHD should only be made by a specialist.

Differential diagnosis These conditions may coexist:

- Learning disability
- Hearing problems
- Epilepsy
- Autism
- Thyroid disease
- Drug ingestion
- Psychological problems (depression, emotional trauma, e.g. divorce)

Management

- Mild/moderate impairment Watchful waiting to see if problems persist, or refer for parent training/education
- Persistent problems/severe impairment Refer to community paediatrics or child and adolescent mental health service for diagnostic assessment. Specialist treatment includes behavioural therapy ± drug therapy (e.g. methylphenidate)
- In all cases Self-help and local support groups can be helpful

Autism spectrum disorder ➔ p. 896

Further information

NICE (2018) Attention deficit hyperactivity disorder: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng87

Information and support for parents and children

British Dyslexia Association ☎ 0333 4054567 🖱 www.bdadyslexia.org.uk
Dyspraxia Foundation ☎ 01462 454 986 🖱 www.dyspraxiafoundation.org.uk
National Attention Deficit Disorder Information and Support Service (ADDISS) ☎ 020 8952 2800 🖱 www.addiss.co.uk

Autism and learning disability

Autism spectrum disorder (ASD) Affects ~1% of the population; ♂ > ♀ (≈4:1). *Risk factors*:

- FH (including FH of psychosis)
- Premature birth (<35wk)
- CNS malfunction/dysfunction
- Valproate in pregnancy
- Intellectual disability
- ADHD
- Genetic disorders, e.g. Down's syndrome, fragile X, tuberous sclerosis, muscular dystrophy, neurofibromatosis

Terminology The *Diagnostic and Statistical Manual of Mental Disorders*, 5th edition (DSM-5), replaced terms '*autistic spectrum disorder*', '*Asperger's syndrome*', '*childhood disintegrative disorder*', and '*pervasive developmental disorder—not otherwise specified (PDD-NOS)*' by the single term 'autism spectrum disorder'. The International Classification of Disease (ICD)-11 is set to follow.

DSM-5 features of ASD Clinically significant impairment in social functioning across multiple contexts that includes:

- **Persistent deficits in social communication/interaction** Including:
  - ↓ social-emotional reciprocity
  - ↓ non-verbal communicative behaviours used for social interaction, and
  - ↓ ability to develop, maintain, and understand relationships
- **Restricted, repetitive behaviour/interests/activities** With ≥2 of:
  - Stereotyped/repetitive motor movements, use of objects, or speech
  - Insistence on sameness, inflexible adherence to routines, or ritualized patterns or non-verbal behaviour
  - Highly restricted, fixated interests that are abnormal in intensity or focus
  - Hyper- or hyporeactivity to sensory input or unusual interests in sensory aspects of the environment (e.g. apparent indifference to pain/temperature, adverse response to specific sounds or textures)

Other developmental disorders (e.g. learning disability/ADHD) and/or mental health problems (e.g. anxiety) often coexist.

Diagnosis^{N} Not apparent at birth; often presents from 18mo–3y with failure of social interaction and lack of speech; may present much later (even as adult) if speech develops. Consider ASD if any concerns about development/behaviour—but be aware of other explanations. *Assess*:

- **Spoken language** e.g. ↓ or absent, monotone, repetitive, only about own interests, rude/inappropriate, one way rather than conversation
- **Response to others** e.g. ↓ responsiveness, misunderstanding of others' intentions, –ve response to requests of others
- **Interacting with others** e.g. ↓ awareness or ↓ tolerance of others entering personal space, plays alone, unaware of socially expected behaviour, difficulty making friends, lack of enjoyment, ↓ eye contact, inappropriate/absent gestures/facial expressions
- **Ideas and imagination** e.g. lack of imagination/creativity
- **Unusual or restricted interests** e.g. dislike of change, over-reaction to stimuli, lack of flexibility, overfocused/unusual interests
- **Rigid and repetitive behaviours** e.g. repetitive stereotyped movements/play, strict adherence to rules
- **Other features** e.g. immature social/emotional skills, lack of common sense, excessive trust, abnormal reactivity to sensory stimuli

Refer to or discuss with the local autism or community paediatric team if ASD is suspected. ❗ Diagnosis of ASD should only be made by a specialist.

Checklists (e.g. Checklist for Autism in Toddlers or CHAT) may be useful to assess possible autism, but do not use to make or exclude a diagnosis.

Management Often severely disabling, lifelong condition requiring substantial support from community services. Most interventions are behavioural and delivered in an educational setting. Medication may be used for specific issues, e.g. melatonin for sleep problems. Be approachable, willing to listen, and an advocate for the family. Give information about self-help and signpost to support organizations and benefits (➔ p. 901).

Learning (intellectual) disability Low intellectual ability (IQ <70), with significant impairment of functioning, and onset in childhood. *Prevalence*: ~2%. May exist alone or with other disabilities. Often noted by a parent first—take concerns seriously. Causes are varied—many are rare. Divide into:

- **Congenital** Genetic (e.g. Down's syndrome, fragile X); metabolic (e.g. congenital hypothyroidism); others (e.g. rubella in pregnancy)
- **Acquired** e.g. trauma, meningitis, birth injury

Outlook Depends on intellectual impairment and coexisting conditions.

- **Mild disability** (80% of learning disability; IQ 50–70) need extra support in school but may go on to lead independent lives
- **Moderate disability** May need special schooling and supervision in later life
- **Profound disability** (often multiple disabilities) need ongoing medical input, special school placement, and life-long care

Management Refer to paediatrics to exclude treatable causes. Then:

- **Communicate** *Explain*: referrals, test results and their implications, the local system and who is responsible for what, where to get more information, including about benefits and housing/schooling. Communicate directly with the child whenever possible; gestures and pictures may help
- **Refer to other community services,** e.g. paediatrics; learning disability service. Ensure follow-up happens and assist with assessment of special needs for schooling, housing, and employment. Provide prescriptions as needed
- **Manage medical problems not related to disability**
- **Promote concordance** With long-term therapy ± education/rehabilitation
- **Reproduction** Offer family planning, pre-conceptual counselling, and/or antenatal diagnosis for parents and patients with learning disability

Learning disability health checks Should be performed annually for adults and children >14y with learning disability—➔ p. 197.

The chronically disabled child ➔ p. 900

Further information

NICE (2011, updated 2017) Autism spectrum disorder in under 19s: recognition, referral and diagnosis. 🖱 www.nice.org.uk/guidance/cg128

NICE (2012, updated 2016) Autism spectrum disorder in adults: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg142

Information and support for parents and children

MENCAP ☎ 0808 808 1111 🖱 www.mencap.org.uk

National Autistic Society of the UK (NAS) ☎ 0808 800 4104 🖱 www.autism.org.uk

Adolescence

Adolescence is characterized by rapid physical development and emotional change. Physical changes start gradually—from ~10y for girls and ~12y for boys—and are complete by the age of ~17y; emotional and social change may take longer. Adjusting to these changes can cause problems:

- Concerns about appearance Some become very concerned about their appearance. They need reassurance, especially if not growing or maturing as quickly as their friends
- Clothes/style Are important to express solidarity with friends and declare independence
- Hormonal changes Lead to body shape, voice, hair, and skin changes, body hair growth, and menstruation. Adjustment can be difficult
- Acne May need treatment—especially if scarring
- Dieting and consumption of junk food Are common. Rarely, eating disorders develop

Consent ➲ p. 48 **Confidentiality** ➲ p. 46

School problems

- School refusal and truancy ➲ p. 888
- Poor school work Emotional problems, e.g. worry about problems at home, often affect school work and concentration; pressure to do well/pass exams may be counterproductive. Exams are important, but advise parents not to let them dominate life or cause unhappiness

Maltreatment ➲ p. 902

Behaviour problems It is normal for teenagers and their parents to complain about each other's behaviour and disagree frequently. Parents often feel they have lost control over their child. Adolescents resent parental restrictions on their freedom—but still want parental guidance. Advise parents to lay down sensible ground rules and stick to them. Evidence suggests children are at greater risk of getting into trouble if their parents do not know where they are—advise teenagers to let their parents know where they are going and parents to ask.

Sexual problems and contraception ➲ p. 746

Trouble with the law ♂ > ♀. Most young people do not break the law—when they do, it usually only happens once. Repeated offending may reflect family culture or may result from unhappiness—always ask about emotional feelings when an adolescent is repeatedly getting into trouble.

Drugs, solvents, and alcohol Most teenagers never use drugs or inhale solvents, a third of 15y-olds experiment, but only 1 in 6 misuse drugs/solvents regularly. Alcohol is the most common drug causing problems for adolescents, but consider the possibility of any form of drug use (➲ p. 162) when parents notice serious, sudden changes in behaviour.

Emotional problems Teenage unhappiness is common and does not necessarily indicate depression (at some point 40% have cried/wanted to get away from everything, and 20% thought life not worth living). However, emotional disorders are often not recognized, even by family/friends. Over-eating, excessive sleepiness, promiscuity, and persistent over-concern with appearance may indicate emotional distress. More obviously, phobias and panic attacks appear.

Rarely, changes in behaviour and mood can mark the beginning of more serious mental health disorders. Bipolar disorder and schizophrenia, as well as more common disorders such as anxiety, may emerge during adolescent years. Refer to the child and adolescent mental health service if concerned.

Normal adolescent behaviour or mental illness? Teenage behavioural problems may be signs of mental illness if:

- They go on for more than a few months
- They do not vary, e.g. persistently low mood in all circumstances
- They are severe, e.g. self-harming behaviour, violence
- There is a significant impact on relationships, school performance, and/or usual activities

Depression in children and young people[N] Response to stress. Distinguish from depressive symptoms occurring as part of other emotional or conduct disorders. Most common in adolescence (♀ > ♂).

Diagnosis Difficult, especially among adolescents. Adolescents often do not communicate well with their parents and have little contact with health professionals—resulting in late diagnosis.

Presenting features

- Unhappiness and/or tearfulness, apathy, boredom, ↓ ability to enjoy life
- Antisocial behaviour—♂ > ♀—especially after bereavement
- ↓ school performance—may admit to poor concentration
- Separation anxiety reappearing in adolescence
- Frequent unexplained illness or undue worries about health
- Self-harm
- Bipolar depressive disorder is rare before puberty, and mania must be present to make a diagnosis

Management Unless a mild episode related to a single precipitating event and no other risk factors for depression, refer for specialist advice. Specialist treatment includes counselling, family therapy, CBT, and drug therapy. (❗ With the exception of fluoxetine, risks of treatment with SSRIs outweigh benefits in children.)

Disorders of puberty ➔ p. 871 **Eating disorders** ➔ p. 992

Further information

NICE (2019) Depression in children and young people: identification and management. 🖱 https://www.nice.org.uk/guidance/ng134

Information and support for parents and children

Brook Advisory Service 🖱 www.brook.org.uk
Childline ☎ 0800 1111 🖱 www.childline.org.uk
Parentline ☎ 0808 800 2222 🖱 www.familylives.org.uk
Phelan T (2016) 1-2-3 *Magic Teen: Communicate, Connect, and guide your teen to adulthood*. Naperville, IL: Sourcebooks. ISBN: 9781492637899.
Young Minds ☎ 0808 802 5544 (parent helpline) 🖱 www.youngminds.org.uk

The chronically ill or disabled child

Chronic disability due to a wide variety of causes affects ~10% of children in the UK.

Effects on the child Vary from child to child, depending on the nature of the disability, personality of the child, and support the child has at home and in the community. Common problems include:

- Physical discomfort—both due to the disability and to painful or embarrassing treatments
- Alterations in the normal pattern of growth and development and/or physical differences may lead to social isolation and ↓ motivation
- Frequent hospitalizations and outpatient visits prevent the child integrating into school or ongoing community activities
- Dependence—the disability may prevent the child reaching his/her own goals and achieving his/her own independence. Many children also realize the additional burden they cause their parents and carers

Effects on the family Vary from family to family depending on financial and/or social support, relationship between parents and other siblings, and many other factors. Stress may cause family break-up, especially when other marital and intra-family problems exist. *Common problems*:

- Grieving for the loss of the 'ideal child'—conditions that affect the appearance of the child particularly affect attachment between parents and child. The grief might take the form of shock, denial, anger, sadness, depression, guilt, or anxiety and may occur any time in the child's development
- Neglected siblings, who may also take on a carer role
- Inconsistent discipline—due to demands placed on the family and sympathy for the child → behaviour problems
- Marginalization of one parent—one parent tends to take on the bulk of the caring activities. There is a danger the other parent starts to feel inadequate and isolated with respect to the care of the child
- Major expense and time commitment—frequently one parent has to give up work to look after a disabled child resulting not only in loss of income, but loss of that parent's independence and opportunities for the future
- Social isolation
- Confusion over the health, benefits, and social services available

Care coordination Inconsistent policies and funding, inadequate access to facilities (including physical barriers to access), and poor communication and coordination between the healthcare, educational, and community support systems causing misery for children with disability and their families. Without coordination of services, care is crisis oriented.

Care coordination requires knowledge about the child's condition, the family, and the community in which they function. In all cases *someone* should be designated responsible for coordinating care—the best person to do that will vary according to circumstances. Regardless of who assists in co-ordination of services, the family and child must be partners in the process.

Prescribing for children ➔ p. 127

Rehabilitation The general principles of rehabilitation for adult patients apply to children too—➔ p. 196 and ➔ p. 554.

Role of the GP

- The GP of any patient with a chronic illness in the community is a team member and may be the key worker who coordinates care
- The GP provides continuity of care, particularly when the child is under the care of several different secondary care teams
- Maintain an open door policy and encourage children and carers to seek help for problems early
- Be flexible with appointments to avoid long waits and allow carers and other family members to attend to their own health needs
- Find out about a child's disease, even if it is rare. It is impossible to plan care without knowledge of course and prognosis, and an easy way to lose a child's confidence if you appear ignorant of their condition
- If progress is slower than expected or stalls, consider other medical problems (e.g. anaemia, infection), behavioural problems, and communication problems (e.g. poor vision/hearing)
- Remember that children with disability are at ↑ risk of maltreatment and that symptoms/signs of maltreatment may be difficult to distinguish from the effects of the child's underlying problem
- Information alone can improve outcome
- Transition from paediatric to adult services can be a particularly difficult time for young people and their families to manage. Planning should begin early, and the GP should be involved. Consideration should be given to allocating the young person a named GP; for some, the GP will take over care, e.g. stable epilepsy, coeliac disease

Support for carers ➔ p. 200

Benefits

- For parents and children ➔ p. 827
- For sickness/disability ➔ p. 108
- For low income ➔ p. 104

Palliative care ➔ p. 904

Information for parents and children

Contact a Family Support and information for families with disabled children (any disability). ☎ 0808 808 3555 🖱 www.cafamily.org.uk

Independent Panel for Special Education Advice (IPSEA) 🖱 www.ipsea.org.uk

National Network of Parent Carer Forums Most areas have parent carer forums who provide information and support locally for children with disability and special educational needs. 🖱 www.nnpcf.org.uk

Together for Short Lives Hospices often provide care for children with life limiting illnesses. ☎ 0808 8088 100 🖱 www.togetherforshortlives.org.uk

Tourism for All Holidays for families with a disabled child. ☎ 0845 124 9971 🖱 www.tourismforall.org.uk

Whizz-Kidz Mobility for non-mobile disabled children. ☎ 020 7233 6600 🖱 www.whizz-kidz.org.uk

Safeguarding children

Children may be mistreated if harm is inflicted upon them or if a responsible person fails to prevent them from coming to harm. In the UK, there are ~58,000 children on child protection registers or the subject of child protection plans.

Classification of child abuse >1 type may occur concurrently

PHYSICAL e.g. hitting, shaking, throwing, burning, suffocating, poisoning, including fabricated or induced illness	EMOTIONAL The child feels worthless, afraid, unloved, or inadequate. Includes age-inappropriate expectations and witnessing domestic violence
NEGLECT Failure to meet the child's basic (including medical) needs; allowing the child to be exposed to danger	SEXUAL Forcing/enticing a child to participate in sexual activities—physical or pornographic

Female genital mutilation (FGM) or forced marriage <16y Both illegal in the UK. If a child reports FGM, or signs of FGM are observed, this MUST be reported to the police. For suspected cases, follow safeguarding procedures. If you suspect a child might be taken abroad for FGM or forced marriage, inform social services and/or the police immediately.

Risk factors ❗ Any child may be a victim of maltreatment.

Parent/carer factors

- Mental illness/learning disability
- Emotional volatility/anger management problems
- Substance/alcohol abuse
- Being abused themselves
- Chronic stress
- Domestic abuse
- Poverty/poor living conditions
- Social isolation
- Lack of engagement with services

Child factors

- History of previous/sibling abuse
- Learning or physical disability

Presentation *Always* have a high index of suspicion. Be aware that gender, culture, and age all affect vulnerability and likelihood of reporting abuse.

Suspect maltreatment if

- The child discloses it
- Story is inconsistent with injuries (⚠ bruising, bleeding, fractures, or other injuries in children not independently mobile are suspicious)
- Characteristic injuries—marks consistent with cigarette burns; scalds (especially if symmetrical or doughnut-shaped on buttocks); finger mark or bite mark bruises; perineal bruising or anogenital injury; linear marks consistent with whipping; buckle or belt marks
- Late presentation or lack of concern about significant injury/illness
- Behaviour of the child is suggestive, e.g. sexually precocious behaviour, persistent abnormal interaction between child and parents
- STI or pregnancy in any child <13y (consider if the child is older)
- The child is persistently smelly/dirty and/or inadequate home environment (including food and hygiene)
- Unexplained stealing/hoarding of food by the child

Consider maltreatment within your differential diagnosis if

- Encopresis, enuresis, or daytime incontinence of urine (➔ p. 892)
- Faltering growth due to lack of provision of adequate diet (➔ p. 850)
- Severe or persistent infestations (e.g. scabies, head lice), oral injuries, or urinary/anogenital symptoms without adequate explanation
- Failure to attend healthcare appointments and/or poor concordance with treatment plans for significant medical/dental conditions
- Unusual/frequent presentation to healthcare professionals
- Injury as a result of inadequate supervision
- Behavioural problems (➔ p. 886)
- Inappropriate dress (e.g. underdressed in cold weather)

⚠ **Immediate action**[N] Do *not* ask leading questions.

- **Listen and observe** Record the history given, any report of maltreatment, the child's appearance and behaviour, any physical signs, and interaction between the child and parent/carer
- **Seek an explanation** From the accompanying adult and (if possible) the child. Record the explanations given. A suspicious explanation is implausible, inadequate, inconsistent (over time or between the child and parent/carer). Cultural practice is not an adequate explanation and never justifies harm
- **Decide** If child maltreatment is likely, a possibility, or can be excluded.

❗ At any stage your level of concern may change and lead you to exclude or suspect maltreatment

Share your concerns With parents and child unless you believe doing so will ↑ risk to the child or where you suspect fabricated or induced illness. Discuss any planned actions. Make a record of your discussions.

If child maltreatment is a possibility Welfare of the child is *paramount*. Check the child's and other family members' records for worrying features; consider asking other practice members if they have concerns. Then consider one or more of the following:

- Seek further information from other agencies (e.g. school, social services) or other health professionals (e.g. health visitor, specialists involved with care)
- Discuss concerns with an experienced colleague, e.g. practice safeguarding lead, locality named professional for safeguarding, or senior paediatrician
- Arrange to review the child at a date appropriate to concern—follow-up if the appointment is cancelled or is not kept

If child maltreatment is likely Refer to children's social care following local child protection procedures. Follow-up telephone referrals in writing in <24h. Actions taken depend on the nature of the suspected maltreatment.

Responding to child protection enquiries Under Section 47 of the Children Act (1989), GPs have a legal obligation to share relevant information whether or not they have consent of the parents.

Further information

GMC (2012, updated 2018) Protecting children and young people: The responsibilities of all doctors. 🖱 www.gmc-uk.org/ethical-guidance/ethical-guidance-for-doctors/protecting-children-and-young-people

NICE (2009, updated 2017) Child maltreatment: when to suspect maltreatment in under 18s. 🖱 www.nice.org.uk/guidance/cg89

NICE (2017) Child abuse and neglect. 🖱 www.nice.org.uk/guidance/ng76

Child death

Palliative care Sadly, some children have illnesses that are incurable. As children enter the terminal stages of their illnesses, the general principles of palliative care apply (➲ p. 1011) but the emotional traumas are often much greater. If possible, engage specialist palliative care services early. Try to maintain continuity of care with as few professionals involved as possible. Provide ongoing support to family members after the child has died.

Sudden infant death syndrome (‘*cot death*’) ~1 in 3000 babies <1y old in the UK are found unexpectedly dead each year. *Peak age*: 1–4mo; ♂ > ♀. Most common in winter months and at night (midnight–9 a.m.). An identifiable cause can be found for 37%—the rest are unexplained. Theories include cardiac arrhythmia and apnoeic attacks.

Risk factors for cot death

- Sleeping with baby on sofa/chair (50× ↑ risk)
- Baby sleeping face down
- Bed sharing
- Loose soft bedding, e.g. duvet
- Smoking (any family member)
- Overheating
- Minor intercurrent illness
- Twin or multiple pregnancy
- Low birthweight

Reducing risk of cot death ➲ p. 827

Management if you are the first person contacted

- Check an ambulance is on its way; go immediately to the scene. Start resuscitation unless clearly inappropriate. Continue until the baby gets to hospital
- If it is clear the baby is dead and cannot be resuscitated, inform the parents sympathetically. Contact the police/coroner and designated paediatrician for unexpected death in childhood. Arrange for the baby to be taken to A&E
- Take a brief history. Record the circumstances of death immediately (e.g. position when found, bedding, vomit). Listen to the parents. Mention the baby by name and do not be afraid to express your sorrow
- If the baby is a twin, the surviving twin is at ↑ risk of cot death and should be admitted to hospital for observation

Management if you learn that a baby has died Provide information to the rapid response team, and attend the initial case discussion if possible. Consider taking part in the scene of death visit to support parents.

Follow-up Review within a few days. There may be some anger directed towards you as often babies have been seen in general practice within a few days or weeks of the death. Do not be defensive or become angry.

- Discuss suppression of lactation if breastfeeding (➲ p. 822)
- Advise parents about likely grief reactions—guilt, anger, ↓ appetite, sleeplessness, hearing the baby cry. Do not forget siblings—they can be deeply affected too. Continue regular review as long as needed and wanted. Be sensitive to anniversaries. Watch for psychiatric illness
- Ensure parents receive written information including details of helplines and self-help organizations. Consider referral for counselling—ideal timing varies
- Refer for specialist obstetric assessment early in the next pregnancy and make sure parents are put in touch with the Care of Next Infant (CONI) scheme (where available). Discuss the use of apnoea alarms

Apnoea alarms Commonly issued to or purchased by parents if they are worried about the risk of cot death. An apnoea alarm cannot be useful unless parents are taught basic life support to a proficient standard. An alarm should not be supplied without this training. There is no evidence that apnoea alarms prevent cot deaths.

Death of a child in other circumstances Death of a child is always difficult. Accidents are the most common cause of death followed by death from childhood cancer. Principles of management used for cot death can be applied.

Child death review

- All deaths of children <18y (excluding stillbirths) are subject to review by a local child death overview panel and should be notified to it
- Unexpected deaths are investigated by a rapid response team, involving police, a senior paediatrician, and other professionals as appropriate
- This rapid response team notifies and gathers information from other professionals involved with the child (including the GP), carries out an initial case discussion, arranges a visit to the scene of death, and then arranges postmortem informed by the investigation
- Further case discussions take place following postmortem. The GP should be invited and sent a report. An appropriate professional is identified to inform the parents of the findings
- The panel arranges support for the parents throughout this procedure—regard this as additional to GP support

Apparent life-threatening events (*brief resolved unexplained events*) Parents may rush a child to A&E or the GP after an episode of pallor ± floppiness. Parents may have attempted mouth-to-mouth resuscitation before the baby starts to respond to them, or may have simply touched the baby or lifted the baby up and received a response. Usually there are no residual symptoms or signs.

Management Difficult. Parents may have misinterpreted normal irregularities in sleep or the child might be unwell and have a physical cause for symptoms, e.g. early stages of a viral infection. Usually parents are very anxious by the time you see the child. Take a careful history and examine the child from top to toe. Treat any cause of symptoms found. If the infant is well, >2mo old, no previous event, the event lasted <1min and there are no concerning features in the history, be as reassuring as possible and play down anxieties.

⚠ Admit the child for observation and further assessment if:

- Any risk factors for cot death
- Clear history of apnoea
- Recurrent episodes
- Aged <2mo
- Attack lasted >1min
- Medical resuscitation was needed
- Difficult social background
- Parent unable to cope

Information and parent support

Child Bereavement Charity ☎ 0800 0288840 🖱 www.childbereavement.org.uk
Child Death Helpline ☎ 0800 282 986 🖱 www.childdeathhelpline.org.uk
Lullaby Trust ☎ 0808 802 6868 (bereavement support) ☎ 0808 802 6869 (information and support) 🖱 www.lullabytrust.org.uk
Together for Short Lives (Hospices and Palliative Care) ☎ 0808 8088 100 🖱 www.togetherforshortlives.org.uk

Chapter 24

Ear, nose, and throat

The mouth *908*
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Otitis media *924*
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The mouth

⚠ Referral for suspected oral cancer[N]

Urgent referral To be seen in <2wk by a specialist oral surgeon:

- Mouth ulceration persisting for >3wk
- Persistent and unexplained lump in the neck

Urgent referral To be seen in <2wk by a dentist:

- Any lump on the lip or in the oral cavity
- Any red or white patch in the oral cavity consistent with erythroplakia or erythroleukoplakia

Dry mouth *Causes*: anxiety, mouth breathing due to a blocked nose, drugs, or Sjögren's syndrome. Look for cause and rectify if possible. Prescribe artificial saliva, e.g. Glandosane®.

Sore mouth Treat the cause. *Consider*:

- Oral thrush
- Aphthous ulcers
- HSV
- Dry mouth
- Trauma (e.g. burn)
- Side effects of chemo- or radiotherapy
- Anaemia
- Hand, foot, and mouth disease (child)
- Gingivitis

Burning mouth syndrome Continuous or intermittent sensation of burning mouth/tongue ± unpleasant taste in mouth without any visible abnormality. ♀ > ♂. Most common around the menopause. Exclude fungal mouth infection. Check FBC, vitamin B_{12}/folate, ferritin, and HbA1c. If no abnormality is found, treat symptomatically. Dietary supplementation with alpha lipoic acid (available OTC—200mg tds for 1mo then od), keeping mouth moist with artificial saliva or sugar-free chewing gum, low-dose amitriptyline (10–25mg od at 5–7 p.m.), and/or CBT may be helpful.

Mouth ulcers Treat the cause. *Consider*:

- Aphthous ulcers
- Trauma, e.g. sharp tooth, false teeth
- Crohn's disease/UC
- Coeliac disease
- Drugs, e.g. steroids, gold
- Reiter's disease
- Behçet's disease
- HSV
- Herpes zoster
- Vincent's angina
- Erythema multiforme
- Self-inflicted, e.g. burns

Leukoplakia Thick, whitish or grey patch usually on the inside of the cheek, tongue, or gum. It is the mouth's reaction to chronic irritation of the mucous membranes. ♂ > ♀. Common in patients who smoke, patients with ill-fitting dentures, and patients who habitually chew on their cheek. Usually benign but may be an early sign of oral cancer. Refer urgently to a dentist for checking[N].

Erythroplakia Reddened area that results when the lining of the mouth thins. The area appears red because the underlying capillaries are more visible. Erythroplakia is a much more ominous predictor of oral cancer than leukoplakia. NICE recommends urgent referral to a dentist to exclude malignancy in *all* cases[N]:

Tongue problems

- **Blue tongue** Central cyanosis—➔ p. 204
- **Dry and furred tongue** Suggests dehydration

- **Geographic tongue** Irregular, smoother, redder patches that change position over time on the dorsum of the tongue. Due to papillae loss. Asymptomatic or causes soreness. Rarely due to vitamin B_{12} deficiency
- **Large tongue** Consider acromegaly, amyloidosis, myxoedema
- **Smooth tongue** Iron, riboflavin, nicotinic acid, vitamin B_{12} or folate deficiency; idiopathic—usually elderly; antibiotic use
- **Sore tongue** Glossitis of anaemia; Crohn's disease, coeliac disease; carcinoma of the tongue; burn; psychogenic causes
- **Strawberry tongue** Yellowish-white tongue coating with the dark red papillae of the tongue projecting through. Associated with scarlet fever although also present in Kawasaki's disease
- **Ulcer** Assume any non-healing ulcer is due to carcinoma of the tongue until proven otherwise. Refer for biopsy to oral surgeon if persists >3wk[N]. Treatment is with surgery or laser ablation ± radiotherapy

Aphthous ulcers Painful white ulcers. Common affecting ≈20%. Usually idiopathic but may be associated with poor health, stress, Crohn's, coeliac, and Behçet's diseases. Most are short-lived. Large ulcers (up to 2cm diameter) can take ~6wk to heal. Most resolve spontaneously. Topical therapies are effective, e.g. hydrocortisone lozenges qds (dissolve in contact with the ulcer). Refer any ulcer not significantly improving >3wk after presentation to exclude malignancy.

Recurrent aphthous stomatitis (RAS) Recurrent crops of ≥1 aphthous ulcer. Usually idiopathic. May be a FH. Can be associated with smoking cessation. Prevalence falls with age. Check FBC, iron, vitamin B_{12}/folate levels, coeliac serology, ± faecal calprotectin (to exclude Crohn's disease). If no underlying cause can be found, treat symptomatically. Consider specialist oral surgery referral if diagnosis is unclear or if recurrent ulcers cause distress.

Oral cancer Usually squamous cell carcinoma. In the UK, lifetime risk is 1 in 75 for ♂ and 1 in 150 for ♀. Incidence is ↑. Major risk factors: smoking (65%); excess alcohol (30%); HPV 16/18 infection linked to oral sex (12%). >90% of cases in the UK are potentially preventable. Lip cancer has good prognosis as it is seen early (>90% 5y survival), but overall survival for mouth cancer is poor (56% 5y survival), mainly due to poor public awareness/late presentation. Usually presents with leukoplakia (white patch), erythroplakia (red patch), or non-healing ulcer (>3wk).

Management Refer suspicious lesions urgently to a dentist or oral surgeon as appropriate for further evaluation[N].

Lichen planus ➲ p. 594 **Erythema multiforme** ➲ p. 569

Oral thrush ➲ p. 610 **Behçet's disease** ➲ p. 493

Herpes simplex virus (HSV) infection (cold sores) ➲ p. 608

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Support and information for patients

Mouth Cancer Foundation ☎ 01924 950 950 🖱 www.mouthcancerfoundation.org

Dental and jaw problems

Gums

- **Bleeding gums** Consider periodontal disease (most common cause); pregnancy; leukaemia, bleeding disorders; scurvy
- **Hypertrophied gums** Associated with phenytoin use
- **Blue line** Along the margin of the teeth—suggests lead poisoning
- **Gum inflammation** Gingivitis—consider immunodeficiency, vitamin C deficiency, DM, leukaemia, drugs, e.g. phenytoin, nifedipine, ciclosporin

Vincent's angina Pharyngeal infection with ulcerative gingivitis. *Management*: phenoxymethylpenicillin 250mg qds po + metronidazole 400mg tds po.

Periodontal disease Disease of the periodontal ligament caused by bacterial plaque and exacerbated by smoking and DM. Occurs in the normal population >30y. Leads to gingivitis, dental abscesses, and tooth loss. Encourage patients to register with a dentist[N]—regular dental care helps prevent dental emergencies and periodontal disease. In the UK, patients experiencing difficulty finding an NHS dentist should telephone their local PCO and ask to be found a dentist.

Toothache Pain/excessive sensitivity to temperature may be a problem with exposed dentine or pulp infection—advise to see a dentist.

Dental abscess Facial swelling and pain related to bacterial infection. Refer to a dentist. Prescribe analgesia if there will be a delay.

Complications of tooth extraction

- **Haemorrhage** Apply pressure by placing wet gauze over socket and get the patient to bite hard for 15min—refer to dentist if not stopping
- **Painful socket and bad taste in mouth** Infection—refer to a dentist. Give analgesia if delay is likely

Loss of tooth through trauma ➔ p. 1090

Cleft lip and palate

Incidence: 1 in 600 live births—half have other abnormalities too (e.g. hypoplastic mandible). Often—though not always—detected at routine antenatal USS. The cleft may be unilateral or bilateral and involve lip and/or palate. Cleft lips are usually repaired in the first few days of life; cleft palates at ~3mo depending on the weight of the baby.

Problems associated with cleft lip and/or palate

- Feeding difficulties with associated poor weight gain
- Aspiration pneumonia
- Hearing problems—particularly glue ear. In some areas children with cleft palate are routinely given grommets at ~18mo. Treat otitis media promptly. Audiology review is important
- Speech problems—refer for speech therapy
- Dental problems—universal with cleft palate. Orthodontic treatment is always required

Temporomandibular joint (TMJ) dysfunction Common disorder affecting ~70% of the population—only 5% seek treatment. Typically presents in early adulthood. ♂:♀ ≈1:4. Aetiology is complex—malocclusion and trauma play a part, and are exacerbated by psychogenic factors.

Assessment Take a careful history noting pain—duration, location, and nature; precipitating/relieving factors; joint noises; restricted jaw function, e.g. locking, poor bite; and non-specific symptoms, e.g. headache, earache, and tinnitus. Examine the head and neck, including the TMJ and mandibular movement. Exclude other disease. Do not X-ray as this yields little useful information—CT/MRI may be requested by specialists.

Patterns of disease 3 patterns of disease:

- Myofacial pain and dysfunction Due to clenching/teeth grinding. Pain is usually worse in the morning. Stress, anxiety, and depression are key features. Poor sleep is common. May have diffuse muscle tenderness
- Internal derangement Articular disc is in an abnormal position and causes restriction of mandibular movement. Pain is usually continuous and exacerbated by jaw movement
- Osteoarthrosis Degeneration of the joint seen on older patients. Crepitus and sounds from the joint occur on jaw movement

Management Reassure and explain the benign nature of the disorder. Suggest simple analgesia, e.g. paracetamol ± ibuprofen. Resting the jaw and avoiding stress may help. Refer those with ongoing problems to oral surgery for review.

Specialist treatment Bite appliance to wear at night helps ~70%. Physiotherapy, behavioural therapy, and exercises also help. Drug treatments include NSAIDs, antidepressants, opioids, and muscle relaxants. Surgery is only occasionally necessary if medical treatment fails.

Dislocated jaw ➔ p. 1095

Fractured mandible ➔ p. 1095

Halitosis Common after sleep. *Short-term halitosis* is associated with acute illness, e.g. tonsillitis, appendicitis (fetor oris), gastroenteritis, diabetic ketoacidosis.

Chronic halitosis Usually caused by bacterial putrefaction of food debris and dental plaque and is related to poor oral hygiene. Associated with gingivitis ± periodontitis. Smoking, alcohol, isosorbide dinitrate, and disulfiram exacerbate the problem. Rarely caused by metabolic disorders, e.g. trimethylaminuria (TMAU or fish odour syndrome). Examine the mouth and recommend a dental check. Advise oral hygiene, e.g. regular brushing of teeth/tongue, dental flossing; smoking cessation; diet advice—avoid garlic, onions, curries; treat any local infection, e.g. gingivitis; mouthwashes, e.g. 0.2% aqueous chlorhexidine gluconate help ↓ dental plaque. Refer if causing distress.

Facial pain ➔ p. 531

Information and support

British Association of Oral and Maxillofacial Surgeons
Temporomandibular joint disorders. 🖱 https://www.baoms.org.uk/patients/conditions/4/jaw_joint_problems
Cleft Lip and Palate Association (CLAPA) ☎ 020 7833 4883 🖱 www.clapa.com

Sore throat and tonsillitis

Each GP sees ≈120 patients/y with sore throat ± tonsillitis (inflammation of the tonsils)—mostly children and young adults. 70% are viral in origin—the rest bacterial (mostly group A β-haemolytic streptococci).

Clinical picture Pain on swallowing; fever; headache; tonsillar exudates; nausea and vomiting; and/or abdominal pain (especially in children due to abdominal lymphadenopathy). ❗ Viral and bacterial infections are indistinguishable clinically but association with coryza, and cough may point to a viral aetiology.

Differential diagnosis Glandular fever especially in young adults with persistent sore throat.

Investigation Not usually undertaken.

- Throat swabs cannot distinguish commensal organisms (40% carry group A β-haemolytic streptococci) from clinical infection, are expensive, and do not give instant results
- Rapid antigen tests give immediate results but have low sensitivity

Management 90% recover in <1wk without treatment. Complications are rare. Advise analgesia/antipyretics (e.g. paracetamol, ibuprofen), ↑ fluids, salt-water gargles, ± analgesic/antiseptic throat spray/lozenges.

Use of antibiotics Antibiotic prescription can be avoided in most, but education about the reasons for not prescribing is vital.

- Benefits Symptom relief (16h less symptoms); slight protection against some complications (e.g. quinsy, otitis media). There is no evidence antibiotics protect against rheumatic fever or acute glomerulonephritis
- Risks Side effects with antibiotic use; ↑ community antibiotic resistance; 'medicalizing' a self-limiting condition—prescribing ↑ faith in antibiotics encouraging re-attendance with sore throat

Give most patients advice and/or a 'delayed prescription' to collect if no better in 2–5d (~70% do not collect). *Choice*: phenoxymethylpenicillin bd/qds for 5–10d or erythromycin bd or qds/clarithromycin bd for 5d. Amoxicillin is more palatable for small children at low risk of Epstein–Barr virus (EBV).

Reasons to give antibiotics immediately[N]

- ≥3 Centor criteria are present or FeverPAIN score ≥4—Box 24.1
- Patient is systemically very unwell
- Symptoms and signs suggestive of serious illness and/or complications (e.g. peritonsillar abscess, peritonsillar cellulitis)
- ↑ risk of serious complications, e.g. heart, lung, renal, liver, or neuromuscular disease, immunosuppression, CF, premature birth

Complications of sore throat All rare:

- Rheumatic fever ➲ p. 246
- Glomerulonephritis ➲ p. 416
- Quinsy (peritonsillar abscess) Usually occurs in adults. *Signs*: unilateral peritonsillar swelling, difficulty speaking, swallowing (even saliva), and opening jaw (trismus). Refer for IV antibiotics ± incision/drainage
- Retropharyngeal abscess Occurs in children. *Signs*: inability to swallow, fever. Refer for IV antibiotics ± incision and drainage

Box 24.1 Centor and FeverPAIN criteria for sore throat

Centor criteria—1 point for each feature:

- Tonsillar exudate
- Tender anterior cervical LNs
- History of fever >38°C
- Absence of cough

FeverPAIN score—1 point for each feature:

- Fever in the previous 24h
- Pus on tonsils
- Attends rapidly (<3d after onset)
- Severely Inflamed tonsils
- No cough/coryza

Score	*Likelihood of Streptococcal infection*	*Score*	*Likelihood of Streptococcal infection*
<3	3–17%	<2	13–18%
≥3	32–56%	2 or 3	34–40%
		≥4	62–65%

Indications for referral to ENT

Urgent referral Any unexplained sore throat for >1mo.[N]

Referral for tonsillectomy

- Recurrent acute tonsillitis Young children have a lot of throat infections; most 'grow out' of the problem. Tonsillectomy is only considered if missing a lot of school/work (e.g. >7 attacks/y for 2y)
- Airway obstruction Very large tonsils causing sleep apnoea
- Chronic tonsillitis >3mo + halitosis
- Recurrent quinsy
- Unilateral tonsillar enlargement To exclude malignancy

⚠ Tonsillectomy carries a small risk of severe haemorrhage. Readmit any patient with bleeding postoperatively for observation.

Glandular fever (infectious mononucleosis) Caused by EBV. Spread by droplet infection and direct contact ('kissing disease'); 4–14d incubation period. Consider in teenagers/young adults presenting with sore throat lasting >1wk. Other symptoms/signs: malaise, fatigue, lymphadenopathy, enlarged spleen, palatal petechiae, ± rash (10–20%). Send blood for FBC (atypical lymphocytes), LFTs (may be abnormal), and EBV testing (Monospot, Paul Bunnell, ± viral titres).

Management Advise rest, fluids, and regular paracetamol, avoid alcohol. Try salt-water gargles or aspirin gargles (only if >16y). Consider a short course of prednisolone for severe symptoms. Treat 2° infection with antibiotics. Advise to avoid contact sports/heavy lifting for >1mo due to risk of splenic rupture. Symptoms may last several months). ⚠ Do not prescribe amoxicillin as it may cause a rash.

Complications 2° infections; rash with amoxicillin; hepatitis; jaundice; pneumonitis; splenic rupture; neurological disturbances (rare).

Tonsillar tumour Most often elderly. *Signs*: unilateral tonsillar swelling, dysphagia, persistent sore throat (>1mo), earache. Refer urgently to ENT for excision biopsy if suspected.

Further information

NICE (2018) Sore throat (acute): antimicrobial prescribing. www.nice.org.uk/guidance/ng84

Hoarseness and stridor

Hoarseness Change in quality of the voice affecting pitch, volume, or resonance. Occurs when vocal cord function is affected by a change in the cords, a neurological or muscular problem. *Causes*:

- Local causes URTI (most common); laryngitis; trauma (shouting, coughing, vomiting, instrumentation); carcinoma; hypothyroidism; acromegaly
- Neurological problems Laryngeal nerve palsy; motor neurone disease; myasthenia gravis; multiple sclerosis
- Muscular problems Muscular dystrophy
- Functional problems

Assessment Weight ↓, dysphagia, or neck lumps add to suspicions of malignancy. Check TFTs in those with weight gain. Indirect laryngoscopy with a mirror can be difficult and give a poor view. ENT departments have thin fibreoptic scopes for direct visualization in outpatients.

⚠ **Referral for suspected laryngeal cancer**N Refer urgently to be seen in <2wk by an ENT surgeon if ≥45y and:

- Persistent, unexplained hoarseness
- Unexplained neck lump

Laryngitis Hoarseness, malaise, ± fever and/or pain on using voice. Usually viral and self-limiting (1–2wk) but occasionally 2° bacterial infection occurs.

Management Advise patients to rest voice, take OTC analgesia (e.g. paracetamol and/or ibuprofen), try steam inhalations. Consider antibiotics if bacterial infection is suspected (e.g. phenoxymethylpenicillin bd/qds for 5–10d)

Vocal cord nodules Can cause hoarseness. Usually precipitated by voice overuse—typically in singers. Visualized at laryngoscopy. Initial treatment is resting the voice; sometimes nodules need surgical removal.

Functional disorders Hysterical paralysis of the vocal cord adductors due to psychological stress. Can cause the voice to ↓ to a whisper or be lost completely. More common among young women.

Management Refer for laryngoscopy to exclude organic cause. Speech therapy and psychological support may help.

Laryngeal carcinoma ♂ > ♀. In the UK, 1 in 175 ♂, and 1 in 800 ♀ develop laryngeal cancer in their lifetime. 93% of cases are potentially preventable. Smoking (relative risk 8.3), excess alcohol (particularly when combined with smoking), and HPV 16/18 infection (risk ↑ 5.4×). The first sign is usually hoarseness followed by stridor, dysphagia, and pain.

Management Refer urgently to ENT if suspectedN. Diagnosis is confirmed with laryngoscopy + biopsy. Treatment is with surgery ± radiotherapy. Early tumours confined to the vocal cord have 90% 5y survival.

Post-laryngectomy problems After laryngectomy patients have a permanent tracheostomy and require practical and psychological support. *Problems include*:

- Excessive secretions
- Recurrent pneumonia
- Stenosis of the tracheostomy site—refer to ENT/oral surgery if severe
- Communication difficulties—ensure referred to speech therapy
- Maintenance of adequate diet—refer to dietician if not maintaining weight and recurrence of tumour has been excluded

Stridor Noise created on inspiration due to narrowing of the larynx or trachea—much more common in children than adults. Treat the cause.

⚠ *Signs of severe airway narrowing*

- Distress
- ↑ respiratory rate
- Pallor and cyanosis
- Use of accessory muscles and tracheal tug

Causes Congenital abnormalities of the larynx; epiglottis; croup (laryngotracheobronchitis); inhaled foreign body; trauma; laryngeal paralysis.

Laryngomalacia (congenital laryngeal stridor) Common in small babies. Due to floppy aryatic folds and the small size of the airway. Stridor becomes more noticeable during sleep, excitement, crying, and with concurrent URTIs. Normally resolves without treatment. Parental concern may necessitate referral.

Croup Common viral infection occurring in epidemics in autumn and spring. Starts with mild fever and runny nose. In younger children (<4y), results in a barking cough and inspiratory stridor. The cough typically starts at night and is exacerbated by crying. Some children have recurrent attacks associated with viral URTI.

Management Steam helps. Steroids can be helpful—give oral dexamethasone 0.15mg/kg, or prednisolone 1–2mg/kg. Admit as a paediatric emergency if there is intercostal recession, cyanosis, or the child's carers are unable to cope.

Acute epiglottitis in children Bacterial infection causing a swollen epiglottis. Can potentially obstruct the airway. Much rarer since introduction of routine *Haemophilus influenza* type b (Hib) immunization. Consider if stridor, drooling, fever, and upright 'leaning-forward' posture.

⚠ If suspected, do not examine the child's throat as this can precipitate complete obstruction.

Management Refer urgently but try to maintain a calm atmosphere to avoid distressing the child. Examination will be undertaken in hospital with full resuscitation facilities on hand. Treatment: IV antibiotics.

Adult epiglottitis Much less common than childhood epiglottitis and less likely to cause complete airway obstruction. Refer for IV antibiotics.

Inhaled foreign body Refer to ENT for assessment.

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Neck lumps and salivary gland problems

Neck lumps are common and can be the first sign of serious underlying pathology. Differential diagnosis—Figure 24.1.

Lymphadenopathy 75% of enlarged LNs are localized and reactive to local pathology; 25% of lymphadenopathy is generalized. *Causes*:

- Benign infective Viral infection, e.g. EBV, CMV, adenovirus, HIV; bacterial infection, e.g. streptococcal sore throat; TB; toxoplasmosis; syphilis
- Benign non-infective Sarcoid; connective tissue disease (e.g. RA, SLE); skin disease (e.g. eczema, psoriasis); drugs (e.g. phenytoin)
- Malignant Lymphoma, CLL, ALL, metastases—both head/neck cancer and lung cancer may present with enlarged cervical LNs

Assessment

- History Duration; symptoms of associated infection, e.g. sore throat; fever; night sweats; shortness of breath; pruritus; weight ↓; and/or alcohol-induced LN pain. Consider taking sexual/drug use history
- Examination Distribution/extent of LNs—localized, soft, tender, mobile LNs suggest acute infective cause; supra-/infraclavicular or firm, non-tender, fixed LNs suggest malignancy; hepatosplenomegaly; if localized, check for potential local causes of lymphadenopathy
- Possible investigations FBC (in <48h if generalized lymphadenopathy[N]); ESR, CRP, or plasma viscosity; HIV test; EBV, and/or CMV serology. Urgent CXR (in <2wk) if respiratory symptoms or ≥40y and supraclavicular and/or persistent cervical lymphadenopathy[N]. Consider autoimmune profile (to exclude SLE and RA) and/or investigations to exclude TB, syphilis, and toxoplasmosis as appropriate

Management Treat the underlying cause as needed. Refer if no cause is found and/or lymphadenopathy persists.

⚠ *Refer urgently*[N]

- Any unexplained neck lump—to ENT/oral surgery to be seen in <2wk
- Unexplained lymphadenopathy—in adults, to haematology to be seen in <2wk; in children, to paediatrics to be seen in <48h

Branchial cyst Arises from embryonic remnants in the neck. Most common in young adults. Presents as a smooth swelling in front of the anterior border of the sternomastoid at the junction of its upper and middle thirds—often during a viral URTI. Fluctuant lump that does not move on swallowing. Treatment is by excision—refer to ENT.

Thyroglossal cyst The thyroid gland develops from the lower portion of the thyroglossal duct. If a portion of this duct remains patent it can form a thyroglossal cyst. Usually presents in young adults (peak age 15–30y) with either a painless, smooth, cystic midline swelling between the isthmus of the thyroid gland and the hyoid cartilage, or just above the hyoid cartilage or, if the cyst is inflamed, a painful, tender lump with localized swelling. *Examination*: the cyst rises as the patient sticks out the tongue. Refer to ENT for excision.

Mumps Table 17.4, ➔ p. 629

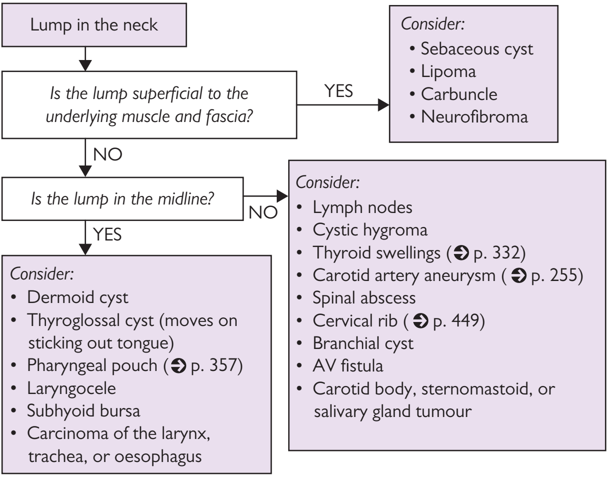

Lump in the neck
Is the lump superficial to the underlying muscle and fascia?
YES
Consider:
• Sebaceous cyst
• Lipoma
• Carbuncle
• Neurofibroma
NO
Is the lump in the midline?
NO
Consider:
• Lymph nodes
• Cystic hygroma
• Thyroid swellings (➔ p. 332)
• Carotid artery aneurysm (➔ p. 255)
• Spinal abscess
• Cervical rib (➔ p. 449)
• Branchial cyst
• AV fistula
• Carotid body, sternomastoid, or salivary gland tumour
YES
Consider:
• Dermoid cyst
• Thyroglossal cyst (moves on sticking out tongue)
• Pharyngeal pouch (➔ p. 357)
• Laryngocele
• Subhyoid bursa
• Carcinoma of the larynx, trachea, or oesophagus

Figure 24.1 Differential diagnosis of neck lumps

Salivary gland tumours Present with a lump/swelling in a salivary gland—80% in the parotid gland. Treated with surgery ± radiotherapy. Refer urgently to ENT/oral surgery if unexplained swelling in the parotid or submandibular gland.

Salivary gland strictures and stones Present with pain/swelling on eating due to obstruction of saliva flow. The gland may appear normal or be tender/swollen. Sometimes stones can be visualized at the salivary duct orifice or felt on bimanual palpation. Predispose to gland infection.

- **Salivary stones** Calculi are most common (80%) in the submandibular duct system; less frequently found in the parotid duct system
- **Strictures** Strictures of the salivary gland duct occur as a complication of a pre-existing calculus, due to mucous plugs or following trauma to the duct wall (e.g. cheek biting)

Refer to ENT/oral surgery for confirmation of diagnosis. Some stones pass spontaneously; most need surgical removal. Strictures can be dilated.

Acute parotiditis Unilateral parotid swelling and pain caused by bacterial infection. Predisposing factors include DM, immunosuppression, local fibrosis following radiotherapy, and autoimmune destruction (e.g. Sjögren's syndrome). Precipitating factors include surgery, dehydration, salivary stones/strictures, and poor oral hygiene. Treat with antibiotics (e.g. co-amoxiclav 500/125mg tds for 1wk) and rehydration. If not settling, consider abscess formation—refer to ENT/oral surgery for drainage.

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Nasal problems

Anosmia

Bilateral anosmia More common than unilateral. *Causes*:

- Local causes URTI, rhinitis, enlarged turbinates, nasal polyps
- Central causes CNS tumours, after head injury, meningitis, hydrocephalus, Kallman's syndrome

Unilateral anosmia One-sided loss of the sense of smell. *Causes*: head injury, frontal lobe lesion.

Taste disturbance Taste is often dependent on smell. Anosmia can also result in taste disturbance. Other causes of taste disturbance include:

- Post-nasal drip
- Drugs—taste disturbance is a side effect of ACE inhibitors
- Burning mouth syndrome (➲ p. 908)—often associated with a bitter, metallic taste in mouth
- Cranial nerve palsy—glossopharyngeal nerve—taste loss on posterior third of the tongue; facial nerve (➲ p. 510)
- Chronic adrenal insufficiency—↑ sensitivity to taste
- Malignancy—taste sensations, e.g. metallic taste with pancreatic cancer

Nasal discharge (rhinorrhoea)

- Clear discharge May be physiological (e.g. due to cold air), due to allergy (e.g. hayfever), or viral (e.g. URTI). The term coryza refers to watery discharge arising from a viral infection
- Clear fluid after trauma Can indicate CSF leak
- Green discharge Indicates active bacterial infection
- Yellow discharge May indicate viral/bacterial infection or allergy
- Persistent blood stained discharge ⚠ Tumour of the nose or postnasal sinus until proven otherwise—refer urgently to ENT

CSF rhinorrhoea Clear fluid dripping from the nose after trauma can indicate a fracture of the roof of the ethmoid labyrinth and CSF leak; it suggests significant trauma. Fluid tests +ve for glucose. Consider referral for head injury assessment. Spontaneous healing of the CSF leak is the norm but, if it persists, refer to neurosurgery for dural closure.

Nasal obstruction Common symptom. Usually of short duration and bilateral. ⚠ Assume persistent unilateral blockage is neoplastic until proven otherwise—refer urgently to ENT.

Causes of nasal obstruction

- Mucosal swelling Coryza, rhinitis (➲ p. 920), iatrogenic, nasal polyps
- Septal deviation Trauma, congenital, e.g. 2° to cleft lip
- Other Tumour, enlarged adenoids, foreign body (➲ p. 1090–1091)

Deviated nasal septum Common in adults—usually 2° to injury. May be associated with external deformity. Nasal blockage is unilateral. Treat mucosal swelling due to rhinitis first as that may be sufficient to control symptoms. If unsuccessful, refer for surgery (submucous resection).

Septal haematoma May occur after injury and causes nasal blockage. Presents as a bilateral soft bulging of the septum. Refer urgently to ENT for evacuation to prevent cartilage destruction.

Septal perforation Can cause bleeding, crusting, and discomfort. *Causes*: trauma, nose picking, cocaine use, postoperative, malignancy. Refer if suspicion of malignancy, otherwise treat symptomatically (e.g. Vaseline® or Naseptin® for crusting)—surgical closure is often not successful.

Post-nasal drip Draining of nasal secretions down the back of the throat. Treat as for chronic sinusitis (➔ p. 920). *Symptoms include*:

- Feeling of mucus in the back of the throat
- Chronic cough—usually worse in the morning and improves in the day
- Morning sore throat
- Nasty taste in the mouth/bad breath

Causes URTI, sinusitis, allergic and/or vasomotor rhinitis, nasal polyps, deviated nasal septum.

Nasal polyps Most common in ♂ aged >40y—associated with asthma, allergic rhinitis, and chronic sinusitis. Consider CF in children <16y. *Symptoms*: nasal blockage; watery discharge; post nasal drip; change in voice; loss of smell; taste disturbance.

Signs Polyps are smooth and pale, usually bilateral, and commonly arise from the middle meatus and middle turbinates. They may completely block the nasal passage. They can be confused with enlarged inferior turbinates but are more mobile and lack sensation.

Management Try medical treatment—steroid nasal drops (e.g. fluticasone) until polyps shrink (maximum 1mo) and then steroid nasal spray to ↓ recurrence. Swab and give antibiotics if purulent nasal discharge. Refer for consideration of polypectomy if medical treatment fails. ❗ Polyps often recur after surgery.

> ⚠ Refer unilateral polyps with an unusual or irregular appearance especially if ulcerating and/or bleeding, for exclusion of malignancy.

Nose bleed/epistaxis ➔ p. 1062

Nasal foreign body ➔ p. 1090–1091

Snoring and sleep apnoea ➔ p. 308

Fractured nose Undisplaced nasal fractures usually heal without intervention. X-ray is unhelpful. Give adequate analgesia. Advise that bruising may be extensive and the nose will feel blocked for 1–2wk.

Associated injuries Consider assessment for head injury (➔ p. 1094). Always look for associated fractures of the zygoma/maxillary bones ('step' deformity in the orbit, dental malocclusion, difficulty opening the jaw, diplopia). Refer urgently to the maxillofacial surgeons if present.

Assessment for permanent deformity Can be difficult at the time of the injury due to soft tissue swelling—reassess 7–10d after injury. Refer to ENT promptly any patient with significant deformity, or if the patient is unhappy with the appearance of the nose. Reduction should ideally take place <3wk after fracture. Deviation of the nasal septum may not be correctable at the time of manipulation and, if symptomatic, will need a later submucous resection.

Sinusitis and rhinitis

Acute sinusitis Infection of ≥1 paranasal sinus (maxillary, frontal, ethmoid, or sphenoid). Usually follows URTI—10% are due to tooth infection. Presents with frontal headache/facial pain (may be difficult to distinguish from toothache)—typically worse on movement/bending ± purulent nasal discharge ± fever.

Management[N] Usually resolves in 7–10d. Advise analgesia (paracetamol ± ibuprofen), fluids ± steam inhalation. *Treatment options*:

- Decongestants—little evidence of effectiveness
- Steroid nasal sprays (e.g. beclometasone 2 puffs to each nostril bd)
- Antibiotics (e.g. amoxicillin 500mg tds)—only if severe symptoms, symptoms persisting >2.5wk, or high risk of serious complications (e.g. CF, immunosuppression)
- Admit as an ENT emergency if suspected orbital cellulitis

Chronic/recurrent sinusitis >3mo of symptoms or >3 episodes of sinusitis in any year. Presents with postnasal drip; frontal headache/facial pain; and/or blocked nose. Associated with nasal polyps (➔ p. 919) and vasomotor rhinitis. Treat as for acute sinusitis. Consider nasal irrigation with saline. Refer to ENT if symptoms are interfering with life.

Rhinitis Inflammation of the nasal mucosa. Affects >1 in 5 people. May be allergic (most children; 1 in 3 adults) or non-allergic (e.g. *vasomotor*—triggered by physical/chemical agents such as cold air, tobacco, or perfumes; *drug induced*). If allergic cause is suspected, ask about allergens: pollen, animals, fungi/moulds, occupational allergens (e.g. flour, latex).

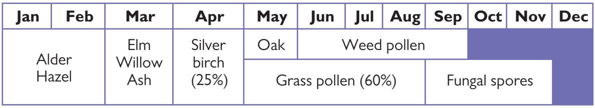
Symptoms Nasal discharge, itching, sneezing, ± nasal blockage/congestion. Symptoms may be seasonal (only certain times of the year) or perennial (all year); intermittent (<4d/wk or <4wk at a time) or persistent. Make an assessment of severity. Patients have moderate/severe symptoms if ≥1 of: troublesome symptoms; abnormal sleep; impairment of daily activities/sport/leisure; problems at work/school. If symptoms are intrusive and difficult to control, refer for allergy testing.

Signs Swollen inferior turbinates; ↓ nasal airway; pale or mauve mucosa; nasal discharge; 'allergic crease' on bridge of nose from persistent rubbing (young sufferers with allergic rhinitis).

Management of allergic rhinitis General measures include ↓ in allergen exposure (➔ p. 281); nasal douching with saline nose drops ± steam inhalation. Drug treatment—Table 24.1.

Desensitization 50—70% success rate. Risk of anaphylaxis is high so provision is limited to specialist centres—refer via an allergy clinic.

Non-allergic rhinitis Treat as for allergic rhinitis—often less effective.



Jan	Feb	Mar	Apr	May	Jun	Jul	Aug	Sep	Oct	Nov	Dec
Alder Hazel		Elm Willow Ash	Silver birch (25%)	Oak	Weed pollen						
				Grass pollen (60%)				Fungal spores			

Figure 24.2 Predominant pollen types at different times of year in the UK

Table 24.1 Drug treatment of allergic rhinitis

Category	Notes
Nasal steroids	Effective if applied properly, and can be used safely long term Take several days to work—try for >2wk before abandoning. Often started at high dose—when symptoms are controlled, dose is ↓ to the minimum that maintains symptom control Choose preparations with minimal systemic absorption if using >1mo (e.g. fluticasone, mometasone). If nasal irritation, sore throat, or nose bleeds switch to a preparation without benzalkonium chloride preservative, e.g. Flixonase® Nasule® or Rhinocort®
Oral steroids	Only rarely needed. *Consider for*: severe nasal obstruction; short-term rescue medication for uncontrolled symptoms; or control of symptoms for important social/work events (e.g. examinations) Use 20–30mg prednisolone PO for 5–7d in combination with nasal steroids. Injected preparations are not recommended
Oral antihistamines	Choose a non-sedative antihistamine, e.g. loratadine 10mg od May be used alone or in combination with nasal steroids. Improve associated symptoms (e.g. conjunctivitis) as well as nasal symptoms
Topical antihistamines	e.g. azelastine nasal drops—useful as a rescue therapy Faster acting than oral antihistamines—onset of action is in <15min
Leukotriene receptor antagonists	e.g. montelukast 10mg od As effective as antihistamines. Useful for patients with concurrent asthma. Combination with antihistamines does not ↑ efficacy
Topical/oral decongestants	e.g. ephedrine nasal drops tds/qds. Effective (drops >>oral preparations) in ↓ nasal congestion Discourage use of nasal drops for >10d as vasoconstriction → mucosal damage → worsening of nasal congestion—a vicious cycle termed *rhinitis medicamentosa*. Not caused by oral preparations
Topical anticholinergics	e.g. ipratropium bromide nasal spray tds—↓ rhinorrhoea but no effect on other nasal symptoms
Topical chromones	e.g. sodium cromoglicate nasal spray—less effective than nasal steroids but may be useful for children or pregnant women wishing to avoid steroids

Hayfever Rhinitis and/or conjunctivitis and/or wheeze due to an allergic reaction to pollen. Occurs at different times depending on the pollen involved (Figure 24.2). When the pollen count is high—keep windows shut (consider pollen filter for the car); wear glasses/sunglasses; avoid grassy spaces. Treat as for allergic rhinitis. Topical chromone eye drops (e.g. nedocromil) may help eye symptoms.

Further information

British Society for Allergy and Clinical Immunology (BSACI) (2017) Guidelines for the management of allergic and non-allergic rhinitis 🖱 https://www.bsaci.org/Guidelines/rhinitis-2nd-edition-guideline

NICE (2017) Sinusitis (acute): antimicrobial prescribing. 🖱 www.nice.org.uk/guidance/ng79

Earache and external ear problems

Earache Ear pain is a common presenting symptom. Think of:

- **Local causes** *Outer ear*: otitis externa; furunculosis; impacted wax; pinna pain (perichondritis); malignant disease of the ear. *Middle ear*: otitis media; barotrauma; myringitis; mastoiditis
- **Referred pain** Trigeminal nerve (dental abscess/caries; impacted molar teeth; TMJ dysfunction); facial nerve (HSV infection; Ramsay Hunt syndrome); vagus nerve (tumours of the piriform fossa, larynx, or post-cricoid area); glossopharyngeal nerve (tonsillitis/quinsy/post-op tonsillectomy; tumour of the base of the tongue or tonsil; neuralgia); cervical nerves C2/3 (cervical spondylosis)

> ⚠ Refer urgently to ENT if persistent, unilateral unexplained pain in the head/neck area associated with otalgia (earache) but normal otoscopy.

Myringitis Inflammation of the tympanic membrane. *Myringitis bullosa* describes painful vesicles on the tympanic membrane associated with mycoplasma or viral URTIs. A similar picture occurs with *Ramsay Hunt syndrome* (➔ p. 512).

Discharge from the ear Otorrhoea is discharge from the ear. Major causes are otitis externa, otitis media, and cholesteatoma. ❗ Always exclude a perforated drum in discharging ears—beware of cholesteatoma. If you cannot visualize the drum—review the patient. Clear fluid leaking from an ear after head injury may suggest a CSF leak. Fluid tests +ve for glucose. This implies a head injury with force—refer to A&E for further assessment.

Otitis externa Inflammation ± infection, of the external ear canal. Common—affecting ~10% at some time. Adults > children. Associated with eczema of the ear canal. *Risk factors include*: swimming, humid environment, narrow ear canal, hearing aid/in-ear headphone use, and mechanical trauma (e.g. cleaning ears with cotton buds, or syringing).

Acute otitis externa <6wk duration. Presents with ear pain (often severe), discharge (may be offensive), and hearing loss ± lymphadenopathy behind/in front of the ear. If the ear canal is not obscured by debris/discharge, it appears red, swollen, and inflamed. Moving the pinna may be painful. ❗ Acute episodes have a tendency to recur.

> ⚠ Immunosuppressed patients and those with diabetes can develop a severe necrotizing form of otitis externa—refer to ENT early.

Chronic otitis externa (>3mo duration)—ongoing discharge from the ear ± hearing loss. Causes canal stenosis and permanent hearing ↓.

Management Although very common, can be difficult to treat. Take a swab if any discharge.

- Advise analgesia (e.g. paracetamol ± ibuprofen)
- Prescribe ear drops—options are: aluminium acetate drops (as effective as antibiotics); and antibiotic and/or steroid drops (e.g. Sofradex®). 💣 If you cannot see the ear drum to ensure that it is intact; use of potentially ototoxic gentamicin ear drops is controversial

- Adding oral antibiotics (e.g. flucloxacillin/erythromycin qds) does not improve outcome—only use if treatment with drops alone has failed or administration of ear drops may be ineffective, e.g. debris within the canal, very swollen canal, uncooperative child

❗ Skin of the pinna adjacent to the ear canal is often affected by eczema. Treat with topical corticosteroid cream/ointment—avoid prolonged use.

If no response after 1 week Try alternative ear drop, e.g. Otosporin® (contains neomycin, hydrocortisone, and an antifungal—polymyxin B) ± oral antibiotics. If swab result is available, prescribe based on the result. Consider gentle syringing to remove infected material. Refer to ENT for aural toilet/advice on further management if no response.

Furunculosis Boil in the ear canal. Presents with severe ear pain—may be exacerbated by moving the tragus or opening the jaw. Exclude DM.
- If no surrounding cellulitis—advise OTC analgesia and application of hot compresses—most will settle. If not settling, prescribe topical antibiotics and steroid drops, e.g. Gentisone HC®, 3 drops qds for 1 wk
- If surrounding cellulitis—prescribe flucloxacillin 250–500mg qds for 7d
- Refer to ENT for incision and drainage if not settling

Foreign bodies in the ear ➔ p. 1090

Ear wax Normal. Becomes a problem only if causes deafness, pain, or other ear-related symptoms. Factors preventing normal extrusion of wax from the ear (e.g. wearing a hearing aid, using cotton buds to clean ears) ↑ the chance of ear wax accumulating.

Ear syringing Indicated if impacted wax causes loss of hearing, discomfort, or tinnitus. Avoid syringing if there is deafness in the other ear or a history of perforation of the ear drum (including grommet), previous mastoid operation, or chronic middle ear disease (e.g. chronic suppurative otitis media, cholesteatoma). If ear syringing is contraindicated, refer to ENT for wax removal under direct vision, e.g. with microsuction.

Perichondritis of the pinna Infection of the pinna due to ear piercing or laceration. If not treated quickly can result in destruction of cartilage and 'cauliflower ear'. Pseudomonas is a common infecting organism so treat with oral ciprofloxacin 500–750mg bd. If not settling, refer as an emergency to ENT.

Chondrodermatitis nodularis helicis (CNH) Caused by pressure on the ear (e.g. against pillow at night, tight headwear). Tender lump, often with overlying scaling/ulceration, on the outer helix of the pinna. Tender to lie on and painful in the cold. Advise relief of pressure ± topical steroid/antibiotic cream. If this fails, cryotherapy or surgical excision is effective. *Differential diagnosis*: SCC, BCC, gouty tophus.

Haematoma of the pinna ➔ p. 1095

Accessory auricle *Incidence*: 1.5/100 live births. Small skin lesion consisting of skin ± cartilage in front of the ear. No treatment is necessary but accessory auricles are often removed for cosmetic reasons.

Bat ears Common congenital abnormality. A fold of the pinna is absent. The child is noted to have protruding ears. Runs in families. Referral for surgery is indicated if the condition is causing psychosocial problems.

Otitis media

Acute suppurative otitis media (OM) Common, acute inflammation of the middle ear. Parental smoking ↑ children's risk of OM—encourage parents to stop smoking. Caused by viral/bacterial infection.

Presentation Ear pain—usually unilateral ± fever/systemic upset. Ear discharge may be associated with relief of pain if there is a spontaneous perforation of the ear drum. *Examination*: red, bulging drum. If perforation has occurred the external canal may be filled with pus obscuring the drum. ❗ If you can't see the drum, review the patient after treatment.

Management[N]

- In 80%, symptoms resolve in ≤4d without treatment. Advise fluids and paracetamol and/or ibuprofen for analgesia and fever control. Symptoms resolve 24h earlier with antibiotics but antibiotics carry the risk of side effects and use ↑ community antibiotic resistance. Consider using a 'delayed' approach—prescribing if symptoms are no better in 4d
- Consider prescribing immediately (e.g. amoxicillin tds) for children with bilateral OM or acute OM with otorrhoea
- Prescribe immediately if very systemically unwell, or at high risk of serious complications because of pre-existing comorbidity, e.g. significant heart, lung, renal, liver, or neuromuscular disease, immunosuppression, CF, young children born prematurely
- If recurrent attacks (>4 episodes in 6mo) or if acute perforation does not heal in <1mo—refer to ENT

Chronic suppurative otitis media Persistent drainage (>1mo) from the ear associated with tympanic membrane perforation and conductive hearing loss. Not usually painful.

- **Central perforation** 'Safe disease'. Treat as for otitis externa (➔ p. 922). Refer to ENT if there is persistent discharge, deafness, vertigo, or earache. Surgery to close the drum may help
- **Attic or marginal perforation** 'Unsafe disease'. May indicate *cholesteatoma*. Refer to ENT for further assessment

Serous/secretory otitis media (glue ear) Non-infected fluid accumulates in the middle ear due to dysfunction/obstruction of the Eustachian tube, e.g. 2° to throat or ear infection, or tonsillar hyperplasia. Most common cause of hearing loss in childhood. More common in children with Down's syndrome or cleft lip/palate. *Symptoms*: deafness ± earache, difficulties with speech/language, ± behavioural problems. *Signs*: dull, concave drum with visible peripheral vessels ± fluid level and/or air bubbles behind the drum.

> *Management in children* Untreated, 75% have no symptoms in <3mo; 5% have bilateral hearing loss persisting >12mo. Glue ear resolves as the child grows older—treatment is aimed at ↓ impact of symptoms until natural resolution. If not resolving, refer to community audiology/ENT depending on local policy. Specialist treatment options include watchful waiting or grommet insertion.

Management in adults Uncommon in adults—usually follows URTI and spontaneously resolves in <6wk. If not resolving or no history of preceding URTI, refer to ENT to exclude post-nasal space tumour.

Grommets Air-conducting tubes inserted through the eardrum to drain the middle ear. Most are extruded spontaneously <9mo after insertion. May need reinsertion if deafness recurs. Patients can swim/bathe but should avoid diving. If discharge from the ear, treat with antibiotic/steroid ear drops ± aural toilet (see Otitis externa, ➔ p. 922).

Mastoiditis Rare complication of acute OM—infection spreads to the mastoid. *Symptoms*: persistent, throbbing earache; creamy, profuse ear discharge; ↑ conductive deafness; fever and general malaise. *Signs*: tenderness ± swelling over the mastoid; ear may stick out; drum is red/bulging or perforated—if the ear drum is normal, it is not mastoiditis. Refer to ENT as an emergency. Treatment is with IV antibiotics.

Cholesteatoma Skin or stratified squamous epithelium growing in the middle ear. Thought to result from formation of a *retraction pocket* in the pars flaccida of the ear drum. Local expansion as the drum desquamates can damage adjacent structures (e.g. facial nerve; semicircular canals—resulting in vertigo). If infected there is an offensive discharge from the ear. *Signs*: perforation of the pars flaccida of the drum with pearly white discharge within it and conductive deafness. Refer to ENT. Treatment is with suction to clear out the cholesteatoma and/or surgery. Following surgery, the ear should be dry and trouble free—if not, refer back to ENT. Lifelong follow-up is required as cholesteatoma recurs.

Tympanosclerosis Thickening and calcification of the tympanic membrane as a result of scarring from recurrent ear infections or after grommet insertion. Usually asymptomatic. No action is needed.

Barotrauma Due to changes in atmospheric pressure (e.g. air travel, diving) in those with poor Eustachian tube function. Presents with a sensation of pressure/pain in one/both ears, hearing loss ± vertigo. There is fluid behind the drum (or perforated drum), haemorrhagic areas in the drum ± conductive hearing ↓. *Usually resolves* spontaneously in 2–3 wk. If perforation has not healed in <1mo refer to ENT.

Prevention Valsalva manoeuvre, yawning, or sucking boiled sweets during flight—particularly during takeoff/landing—encourages the Eustachian tube to open to allow pressure to equalize. Decongestants may help, e.g. pseudoephedrine 120mg 30min prior to flight. Patients with otitis media should not fly.

Further information

NICE (2018) Otitis media (acute): antimicrobial prescribing. 🖱 www.nice.org.uk/guidance/ng91

Deafness

Congenital deafness Usually detected on neonatal screening (➲ p. 836). *Causes*

- Genetic (50%)
- Intrauterine infection, e.g. rubella
- Drugs given in pregnancy, e.g. streptomycin
- Birth asphyxia
- Meningitis
- Severe neonatal jaundice

Childhood deafness Temporary deafness is common due to middle ear infections but permanent deafness rare (1–2/1000). ↓ hearing is often noticed by parents or teachers—take concerns seriously and refer for assessment. Deafness causes long-term speech, language ± behavioural problems and early intervention makes a difference.

Management History, examination, assess development (including speech and language), consider referral for audiology or to ENT. *Causes*:

- If no earache Bilateral glue ear (➲ p. 924); impacted wax; hereditary cause; sequel of meningitis, head injury, or birth complications
- If earache Acute otitis media (➲ p. 924); impacted wax

Adult deafness Common and debilitating, leading to isolation and depression. Presentation tends to be late. *Causes*: Table 24.2.

Presentation Usually hearing loss develops insidiously with increasing problems understanding others when there is background noise. Tinnitus may be the presenting problem.

⚠ *Refer to ENT if:*

- Conductive deafness of unknown cause
- Sudden deafness if no wax visible
- Asymmetrical deafness—refer urgently to ENT to exclude rare dangerous diagnoses, e.g. acoustic neuroma, cholesteatoma

Useful screening questions

- Do other people mumble a lot?
- Do you find yourself frequently saying 'pardon'?
- Does the family say the TV is too loud?
- Do you miss hearing the doorbell or phone?
- Do you occasionally get the wrong end of the stick in a conversation?

Management Examine the drum; exclude wax; consider postnasal space tumour. If no self-limiting cause is found, refer for a hearing test to quantify hearing loss and assess suitability for hearing aid.

Presbycusis Very common. Causes bilateral symmetrical sensorineural deafness in the over 50s. Deafness is gradual in onset. High frequencies are more severely affected, so speech discrimination—particularly of high-pitched voices—is lost first. Examination is normal. Refer for an audiogram to confirm diagnosis and then for a hearing aid if appropriate.

Otosclerosis Bilateral conductive deafness due to adherence of the stapes footplate to the bone around the oval window. May be FH (50%). If deteriorates in pregnancy, avoid prescribing combined contraceptives. Refer to ENT for assessment to replace the stapes with an implant.

Table 24.2 Causes of adult deafness

Conductive deafness	Sensorineural deafness
Impacted wax (➲ p. 923)	Presbycusis
Debris/foreign body in the ear canal	Noise-induced deafness
Perforation of the ear drum	Infections (measles, meningitis)
Middle ear effusion (glue ear)	Ménière's syndrome (➲ p. 929)
Otosclerosis	Drugs, e.g. aminoglycosides, furosemide
	Acoustic neuroma

Noise-induced deafness Caused by exposure to noise >85dB. May occur in work, or non-work settings (e.g. firearm sports). Immediate indications are ringing in the ears/muffling of hearing after exposure. Refer to audiology. Avoid further excessive noise exposure. Hearing aids may help. If employment-related may be eligible for compensation (➲ p. 91; war veterans—➲ p. 107). Employees should be protected from noise and provided with ear protection if working in noisy environments.

Acoustic neuroma Slow-growing neurofibroma arising from the acoustic nerve. *Symptoms*: unilateral sensorineural deafness, tinnitus, ± facial palsy. *Management*: Refer to ENT. Treatment is surgical.

⚠ Refer urgently to ENT to exclude acoustic neuroma if unilateral or asymmetrical sensorineural deafness.

Benefits for deaf people ➲ p. 108

Hearing aids Can help anyone with reduced hearing, but they never restore perfect hearing. May be:

- Body-worn, behind-the-ear, or in-the-ear aids
- Analogue or digital aids Most traditional aids are analogue aids and amplify all sounds including background noise. Digital aids can be programmed to filter out background noise and customized to the individual's pattern of hearing loss; whistle less; and can have different settings for different sound environments, e.g. TV, crowded rooms
- Bone conduction aids For those with conductive hearing loss or if unable to wear conventional aids due to surgery/malformation
- CROS/BiCROS aids For those with unilateral complete deafness. CROS hearing aids pick up sound from the side with no hearing and feed it to the better ear. BiCROS aids amplify sound from both sides and feed it into the ear that has some hearing

Cochlear implants Benefit patients of any age with profound bilateral sensorineural hearing loss. 2 components—one external (worn behind the ear) and the other internal (surgically implanted). Intense speech therapy is needed for several years to interpret signals from the implant.

Information and support for deaf patients and their carers

Action on Hearing Loss ☎ 0808 808 0123 *Text phone*: 0808 808 9000 🖱 www.actiononhearingloss.org.uk
British Deaf Association 🖱 www.bda.org.uk
National Deaf Children's Society ☎ 0808 800 8880 🖱 www.ndcs.org.uk

Tinnitus and vertigo

Dizziness and giddiness ➜ p. 523

Tinnitus Ringing or buzzing heard in the ears or head. Occasional tinnitus is common (15% of population) but 2% are severely affected. Patients with tinnitus that interferes with daily life and sleep are prone to depression. *Cause*: often unknown. May accompany hearing loss, or be due to noise exposure, head injury, Ménière's disease, anaemia, ↑ BP, or drugs (loop diuretics, tricyclics, aminoglycosides, aspirin, NSAIDs).

Management Reassure patients that there is no sinister cause. Refer to audiology for a hearing aid if there is deafness. Drugs are not helpful but look for and treat associated depression. Psychological support is important—consider referral to a hearing therapist and/or support group (e.g. Tinnitus Association). Masking with background music/radio or an aid that produces white noise (available via ENT) can help. Surgical sectioning of the cochlear nerve is a last resort → deafness.

Indications for referral to ENT

- **Objective tinnitus** Noise can be heard by an observer—rare and may be due to vascular malformations or TMJ problems
- **Unilateral tinnitus** Especially if associated with deafness—refer to exclude acoustic neuroma

Vertigo An illusion that the surroundings are spinning. Ask about duration and frequency, associated nausea, deafness and tinnitus, and recent viral symptoms. *Causes*:

- **Episodic vertigo lasting a few seconds or minutes** Commonly due to benign positional vertigo
- **Episodic vertigo lasting minutes to hours** Consider Ménière's disease
- **Prolonged vertigo (>24h)** Peripheral lesion, e.g. viral labyrinthitis or trauma, or a central lesion (usually associated with other signs), e.g. multiple sclerosis, stroke, tumour

Examination

- Look for neurological signs especially cerebellar signs, cranial nerve lesions, and Romberg's sign
- Assess BP, nystagmus, ear drums, and hearing
- Hallpike manoeuvre:
  - Move the patient quickly from a sitting position to a lying supine position with head turned to one side and extended over the end of the bed—look for nystagmus and ask about vertigo
  - Repeat with the head turned to the other side

Nystagmus Involuntary, oscillatory eye movements—can be congenital or due to labyrinthine or visual system problems. Refer all cases, unless associated with self-limiting labyrinthitis, for assessment.

⚠ Sudden attacks of vertigo can be dangerous. Consider risks of swimming, dangerous machinery, and ladders. Advise patients to stop driving and inform the DVLA—if group 1 licence can resume once symptoms are controlled; group 2 licences are restored if symptom free for >1y.

Benign positional vertigo Recurrent attacks of sudden-onset vertigo lasting only a few seconds or minutes. Occur with sudden changes in posture. Common after head injury or viral illness. Possibly caused by otoliths in the labyrinth. Diagnosis is based on history and a +ve Hallpike test. Normal tympanic membrane.

Management Usually self-limiting (few weeks)—although may continue intermittently for years. Reassure. Labyrinthine sedatives are not helpful. Teach the patient to minimize symptoms by sitting and lying in stages. Habituation may occur by maintaining the trigger position until vertigo settles. If not settling, perform (or refer to ENT for) Epley's manoeuvre^{C} (rapid repositioning of head to move otoliths out of the labyrinth), and/or refer to physiotherapy for exercises/vestibular rehabilitation.

Viral labyrinthitis Usually follows a viral URTI.

- Symptoms/signs Sudden onset of vertigo, prostration, nausea and vomiting, no associated loss of hearing, normal tympanic membrane
- Treatment Labyrinthine sedatives, e.g. cyclizine or prochlorperazine
- Natural history Usually resolves in 2–3wk—if persists >6wk refer

Ménière's syndrome Overdiagnosed in patients with recurrent vertigo and deafness. It is a complex of symptoms including clustering of attacks of vertigo and nausea, tinnitus, a sense of fullness in the ear, and sensorineural deafness which may be progressive. *Aetiology*: idiopathic dilation of endolymphatic spaces.

Management

- Refer all suspected cases to ENT or neurology to confirm diagnosis
- Provide information and advise about support organizations
- Treat acute attacks with labyrinthine sedatives, e.g. cyclizine or prochlorperazine. Consider buccal/rectal routes of administration if vomiting. Do not use long term
- Encourage patients to mobilize after an acute attack
- Betahistine taken regularly may help in some patients, as may thiazide diuretics, a low-salt diet, vestibular rehabilitation, tinnitus maskers, and/or hearing aids
- There is some indication that stress may precipitate attacks
- Look out for and treat concurrent anxiety and depression
- Labyrinthectomy is a last resort and can help vertigo but results in deafness on that side

Vertebrobasilar insufficiency Common in older patients. History of dizziness on extension and rotation of the neck. Normal tympanic membranes. May have associated cervical spondylosis and neck pain. Provide lifestyle advice. 💣 Some advocate use of a cervical collar.

Information and support for patients

Action on Hearing Loss ☎ 0808 808 0123 *Text phone*: 0808 808 9000 🖱 www.actiononhearingloss.org.uk
British Tinnitus Association ☎ 0800 018 0527 🖱 www.tinnitus.org.uk
Ménière's Society ☎ 01306 876883 🖱 www.menieres.org.uk

Chapter 25

Ophthalmology

'The eye is the window of the mind'
Richard II, William Shakespeare (1564–1616)

Assessment of the eye *932*
Eye trauma *936*
Eye pain, papilloedema, and orbital disease *938*
Lid disease *940*
Blepharitis and tear duct problems *942*
The red eye and conjunctivitis *944*
Corneal, scleral, and uveal disease *946*
Visual field loss and blindness *948*
Sudden loss of vision in one eye *950*
Gradual loss of vision *952*
Glaucoma *954*
Cataract *956*
Refraction errors and squint *958*
Drugs for the eye and contact lenses *960*

❗ **Community-based optometrists** are a valuable resource available to GPs to help differentiate eye conditions. In addition to slit lamps, many community optometrists now have optical coherence tomography (OCT) scanners, which are particularly helpful for diagnosing retinal and optic nerve disease.

Getting an optometrist's opinion can direct appropriate referrals, assist in setting the appropriate priority level for referral, and prevent unnecessary referrals. However, patients with rapid sight loss should be referred direct to eye casualty.

Assessment of the eye

History Ask about pain, redness, watering, change in appearance of the eye, altered vision, and if the problem is unilateral or bilateral. Distinguish between blurred and double vision. Enquire about trauma, previous similar episodes, systemic illness, and eye disease in the family. If using medication, take a drug history (including eye drops).

Visual acuity Test and record the central (macular) vision of each eye separately for near and distance vision with glasses on. Distance visual acuity is tested at 6m from the vision chart. Cover the non-tested eye. On a full-size Snellen chart, the '6' line indicates what a normal person sees at 6m; the '60' line indicates what a normal person sees at 60m—Figure 25.3—➔ p. 935. Record as 6/'the line read'. If below 6/6 use a pin hole to improve refraction. Cover the non-test eye carefully. Near vision can be checked using a near-vision testing card (Figure 25.2—➔ p. 934) or newspaper held at a distance of 35cm from the eyes.

Examination

- Eyelids should be symmetrical. Check skin around the lids, eyelash position, and for inflammation, crusting, or swelling of the lid/lid margin
- Use a bright light to examine the eye surface—it should be bright and shiny. In infected eye/contact lens wearer, look carefully for white spots on the cornea. Use a fluorescein stain and examine the surface of the eye with a cobalt blue light if any indication of corneal damage
- Note any redness—if conjunctiva is diffusely inflamed, conjunctivitis is likely; a duskier redness around the margin of the cornea (ciliary congestion) suggests disease of the cornea, iris, or deeper parts of the eye

Direct ophthalmoscopy Takes practice. Darken the room and ensure you have good batteries in your ophthalmoscope.

- Check the red reflex (opacities within the eye appear as a shadow)
- Examine the disc—put your hand on the patient's forehead and support the upper lid with your thumb; use your right eye for the patient's right eye and vice versa. Look for shape, colour, and size of the cup
- Follow each of the 4 main vessels to the periphery
- Examine the macula by asking the patient to look directly at the light, and peripheral retina by asking the patient to look up/down, right/left

❗ Dilating the pupils with a short-acting mydriatic (e.g. 0.5–1% tropicamide) makes examination easier; warn patients that drops may sting and they may have temporarily blurred vision and should not drive home. Rarely, dilating the pupil may precipitate acute angle-closure glaucoma.

Visual fields Test peripheral vision by sitting in front of the patient and comparing their visual field to your own (1 eye at a time)—the most basic test is to check the patient can see hand movement in each of the 4 quadrants. Refer for formal tests. Visual field defects—➔ p. 948.

Eye movements If the patient complains of double vision, move an object to the 9 positions of gaze (Figure 25.1). Ask the patient to tell you in which direction the double vision increases.

Pupils Should be round, central, of equal size, and respond equally to light and accommodation. *Pupil abnormalities*:

- **Horner's syndrome** ➔ p. 270
- **Afferent defect (fixed dilated pupil)** *Causes*: trauma (e.g. blow to the iris), mydriatic drops, acute glaucoma, 3rd nerve palsy
- **Relative afferent pupillary defect (RAPD)** Pupils are the same size but there is a ↓ in constriction response to light in the affected eye. Shine a bright light in the better eye for 3s, then move it rapidly to the affected eye. If the afferent pathway is ↓, the first pupil movement is dilation not constriction. *Causes*: optic neuritis, retinal disease
- **Argyll Robertson pupils** Bilateral, small, irregular pupils with no light response, but normal accommodation. Occurs in patients with DM or neurosyphilis
- **Holmes–Adie pupil** Accommodation is partially paralysed causing blurring of near vision, slight pupil dilation, and a very slow pupil response to light and accommodation (minutes). Occurs unilaterally in young adults and is not associated with serious neurological disease

❗ These Snellen charts are for illustration purposes only. Lines on a full-sized chart are read from a distance of 6m with distance glasses if worn. Read from the top of the chart to the bottom. *Interpretation*:

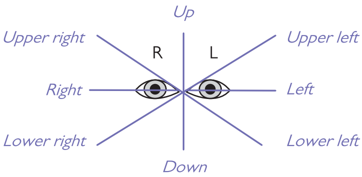


Figure 25.1 The 9 positions of gaze (straight ahead is one position)

Table 25.1 Eye referrals

Emergency referral (to A&E or emergency eye clinic)	• Sudden loss of vision • Acute glaucoma • Perforating injury, intraocular foreign body • Chemical burns • Retinal detachment	• Corneal ulcer • Sudden onset of diplopia or squint + headache • Temporal arteritis with visual symptoms ➔ p. 498
Same-day (<24h)	• Hyphaema or vitreous haemorrhage • Corneal foreign bodies or abrasions	• Orbital fracture • Sudden onset of ocular inflammation, e.g. iritis or ophthalmic herpes zoster
Urgent (<2wk)	• Central visual loss • Chronic glaucoma with pressure >35mmHg	• Sinister 'floaters' • Flashing lights without a field defect
Routine referral	• Gradual loss of vision • Chronic glaucoma (unless pressure >35mmHg) • Chronic red eye conditions	• Painless diplopia or squint • Chalazion/stye/cyst • Ptosis[a]

[a] Urgent if sinister cause suspected.

N.48

She waved

N.36

Faces the sun

N.24

Painting the rainbow

N.18

Life was like a flying dogfish

N.14

Quietly a storm drove purple ducks
across the road. The chimney top

N.12

Glowed in the dusk and my sister let her
biscuit fall through ashes. September was

N.10

In drizzling mood when hedgehogs threw
pinecones in the dark. Squirrels played classical
music.

N.8

We won a feather duster by encouraging Jessica to bake an
enormous apple pie and pirouette between the tables.

N.6

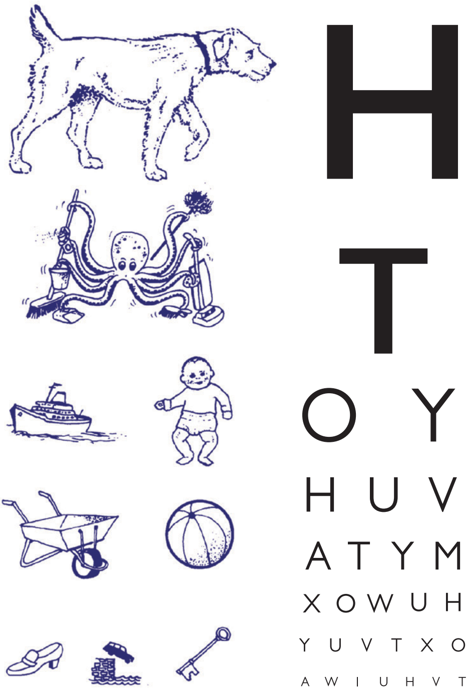
Queuing had never appealed to the young porcupines but swimming held great drama for
the blue and pink ostrich.

N.5

Delight was exceeding the pleasures of everyday ambulation and breaking the pattern of a melancholy
existence to see the trees.

Figure 25.2 Near-vision testing card—reading types are read at 35cm with reading glasses if used. Test each eye separately

Reproduced from Collier J et al., *Oxford Handbook of Clinical Specialties* (2013), with permission from Oxford University Press.



❗ These Snellen charts are for illustration purposes only. Lines on a full-sized chart are read from a distance of 6m with distance glasses if worn. Read from the top of the chart to the bottom. *Interpretation:*

Able to read down to the line labelled:	*6*	Normal vision	6/6
	9	Can see at 6m what a normal person can see at 9m	6/9
	12	Can see at 6m what a normal person can see at 12m	6/12
	18	Can see at 6m what a normal person can see at 18m	6/18
	36	Can see at 6m what a normal person can see at 36m	6/36
	60	Can see at 6m what a normal person can see at 60m	6/60
Counts fingers		Counts fingers held at 0.5m distance	CF
Hand movement		Perceives hand moving at 0.25m distance	HM
Perceives light		Can see a torchlight when shone into the eye	PL
No perceived light		Blind	No PL

Figure 25.3 Snellen charts

Reproduced from Collier J et al., *Oxford Handbook of Clinical Specialties* (2013), with permission from Oxford University Press.

Eye trauma

In all cases

- Take a careful history; establish the nature of the trauma (i.e. what hit the eye and with what force?)
- Measure acuity and examine both eyes carefully, recording your findings
- If the patient is unable to open the injured eye, try to instil local anaesthetic drops and then examine—if unable to do so, refer to eye casualty for assessment
- Encourage accident prevention, e.g. wearing protective goggles

Corneal abrasion

- Take a careful history to exclude high-speed particles, (e.g. from strimmer etc.) that could cause penetrating injury
- Abrasions may cause severe pain—if so, apply a few drops of local anaesthetic (e.g. proxymetacaine 0.5%) before examining
- Use fluorescein stain, with cobalt blue light illumination to detect abrasion—stains green (Figure 25.4)
- If the abrasion is vertical, ensure no foreign body is left in the eye by everting the upper lid
- Abrasions normally heal in <48h; advise chloramphenicol 0.5% eye drops qds until healing is complete
- Eye padding is not needed except to protect the eye after a local anaesthetic

Superficial foreign bodies Cause discomfort, a 'foreign body sensation', and watering. They can be difficult to see so examine very carefully (Figure 25.5), including everting the eyelids. The foreign body sensation may come from an abrasion.

Management

- If metal or a penetrating injury is suspected, refer to eye casualty
- Superficial foreign bodies can be removed with a corner of clean card after instilling local anaesthetic. If that fails or if you are not confident, refer to eye casualty
- After removal, treat with topical antibiotics, e.g. chloramphenicol 0.5% drops 2-hourly for 3d then qds for 4d
- If left >12h, a rust ring may form around a metal foreign body—refer to eye casualty for removal

Photokeratitis (arc eye) Due to corneal epithelial damage as a result of exposure to UV light. Seen in welders, sunbed users, skiers, mountaineers, and sailors who do not use adequate eye protection. Symptoms include severe eye pain, watering, and blepharospasm a few hours after exposure.

Management Pad the eye and give analgesics and cyclopentolate 1% eye drops bd (causes pupil dilation). Recovery should occur in <24h—if not, refer. Advise on suitable protective wear for future exposure.

Blunt injury Caused by fists, squash balls, etc. The result may be anything from a 'black eye' to globe rupture. Globe rupture is usually obvious with a wound and severely ↓ vision. More minor injuries include subconjunctival haemorrhage (➔ p. 945) or corneal abrasion.

Refer urgently if

- Visual acuity is affected
- Double vision
- Lacerated conjunctiva
- *Hyphaema*—blood in the anterior chamber
- Unable to see posterior limit of a subconjunctival haemorrhage—may indicate orbital fracture
- Persistent pupil dilation—usually recovers spontaneously but may indicate a torn iris
- Any signs of retinal damage (oedema, choroidal rupture), *or*
- You cannot assess the eye, e.g. if lid swelling/pain prevents examination

'Blow-out' fracture of the orbit Uncommon fracture due to blunt trauma to the eye (e.g. squash ball injury). Can present with blurred or double vision and pain on moving the eye. *Signs*: enophthalmos (often masked by swelling), infraorbital nerve loss, and inability to look upwards due to trapping of inferior rectus muscle. Refer for X-ray and assessment of eye trauma via A&E.

Penetrating wounds Refer urgently to eye casualty if penetrating injury is a possibility, i.e. history of flying object or working with hammers, drills, lathes, or chisels where a metal fragment may fly off. X-ray/CT scan can confirm diagnosis and help locate the foreign body. Symptoms/signs:

- Wound may be tiny
- Eye is painful and waters
- Vision may initially be normal, or may be very poor, depending on the size of the foreign body
- Photophobia, hyphaema, and/or pupil distortion

⚠ *Do not remove large foreign bodies (dart or knife)* Support the object with padding while transferring the patient supine to eye casualty or A&E. Cover the other eye to prevent damage from conjugate movement.

Chemical burns Can cause great damage—particularly alkali injuries. Use topical anaesthetic (e.g. proxymetacaine 0.5%) before examining. Hold the lids open, brush out any powder, and irrigate with large amounts (1–2L) of clean saline or water immediately. Do not try to neutralize the acid or alkali. Refer urgently to eye casualty.

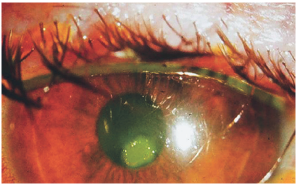


Figure 25.4 Corneal abrasion: stained with fluorescein appears green

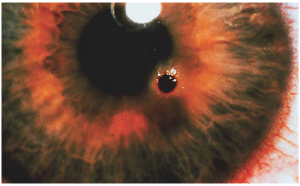


Figure 25.5 Corneal foreign body

Eye pain, papilloedema, and orbital disease

Eye pain *Consider*:
- **Painful conditions** Corneal foreign body, keratitis, iritis, scleritis, acute glaucoma, ophthalmic shingles, arc eye
- **Gritty eye discomfort** Conjunctivitis, entropion, trichiasis, dry eye, episcleritis
- **Pain on moving the eye** Optic neuritis
- **Referred pain** Tension-type headache, migraine, refractive error, trigeminal neuralgia, ophthalmic shingles, giant cell arteritis, ocular muscle imbalance, ↑ ICP
- **Photophobia** Painful vision in normal light. One of the 3 principal features of meningism associated with meningitis; discomfort in the light can also be due to eye disease (e.g. conjunctivitis) and migraine

Disc swelling and papilloedema (Figure 25.6). *Causes*:
- Intracranial SOL
- Encephalitis
- SAH
- Benign intracranial hypertension
- Malignant hypertension
- Optic neuritis
- Disc infiltration, e.g. leukaemia
- Ischaemic optic neuropathy
- Retinal venous obstruction
- Metabolic causes, e.g. hypocalcaemia

⚠ Refer suspected disc swelling for same-day specialist medical opinion.

Swelling around the eyes Oedema around the eyes gives the face a bloated appearance. Swollen eyelids may partially close the eyes. In severe cases the whole face becomes oedematous. Associated with nephrotic syndrome, allergic reactions (e.g. pollen, dust, or insect bites), angio-oedema, and periorbital cellulitis.

Exophthalmos The eyes protrude from the orbit and thus have a staring appearance. Stand at the same level as the patient and look at the patient's eyes. There should be no white of the sclera visible below the iris. If the eye is pushed forward, as in exophthalmos, white sclera is seen below the iris and the patient can look upwards without moving his/her eyebrows (distinguishes from lid retraction).
- **Bilateral** Caused by thyroid eye disease (➔ p. 335)
- **Unilateral** Caused by thyroid eye disease (➔ p. 335), orbital disease (e.g. tumours, cellulitis); vascular disease (e.g. cavernous sinus thrombosis, carotid-cavernous fistula); sinus disease (e.g. tumour)

Microphthalmos 1 in 1000 live births. Small eyes. Associated with Down's syndrome and other genetic abnormalities.

Orbital inflammation

Preseptal cellulitis Infections of the upper lid may cause significant swelling and redness around the eye. Typically affects children following mild trauma. The eye is unaffected—infection is localized to skin and superficial tissues.

Treat as localized cellulitis with oral antibiotics (e.g. flucloxacillin). Monitor carefully as can progress to orbital cellulitis.

Orbital cellulitis Typically due to spread of infection from the paranasal sinuses. Usually presents with pain, double/blurred vision, and general malaise. *Signs*: fever, eyelid swelling, proptosis, and inability to move the eye. Severe cases can lead to septicaemia, meningitis, and cavernous sinus thromboses. If suspected, refer immediately to ophthalmology for IV antibiotics/surgical drainage.

Orbital tumours The eye is in a confined space within the orbit. Any ↑ in mass pushes the eye forwards. *Symptoms/signs*:

- Unilateral proptosis is tumour until proven otherwise
- Orbital pain—especially in rapidly growing malignant tumours
- Lid swelling/distortion
- Limitation of eye movements ± diplopia
- ↓ visual acuity if involvement of optic nerve, retina, or vascular supply

> ⚠ If suspected—refer for urgent/same-day ophthalmology opinion.

Tumours may be

- Primary Benign or malignant—any orbital structure may be involved, e.g. lacrimal gland (carcinoma or adenoma); retina (retinoblastoma in children, melanoma); optic nerve (neurofibroma, astrocytoma, meningioma); lymphoid tissue (lymphoma); connective tissue (rhabdomyosarcoma—rapid growing causing proptosis, ocular inflammation, and poor vision due to optic nerve involvement)
- Due to spread from adjacent structures e.g. post-nasal space tumour
- Due to blood-borne metastases e.g. breast, leukaemia, neuroblastoma, Ewing's sarcoma

Information for patients and carers

Eye Health UK 🖱 www.eyecaretrust.org.uk
Microphthalmia, Anophthalmia and Coloboma Support (MACS)
☎ 0800 169 8088 🖱 www.macs.org.uk

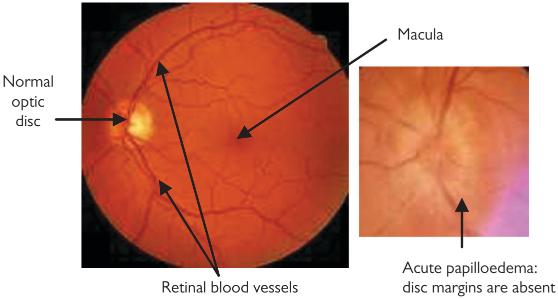


Figure 25.6 The normal retina (left) and papilloedema (right)

Lid disease

Ingrowing lashes (trichiasis) Causes an irritable foreign body feeling in the eye ± recurrent infection. In severe cases, the ingrowing lashes may damage the cornea. Refer to ophthalmology.

Loss of eyelashes (madarosis) Usually due to blepharitis (➔ p. 942) in which case the condition is bilateral and associated with other symptoms/signs of blepharitis. Other causes include plucking/rubbing (may be unilateral or bilateral), alopecia areata, and discoid lupus (scarring madarosis). Sometimes no cause is found. Treat the cause if possible.

Depigmentation of the eyelashes (poliosis) Vitiligo can affect the eyelids. There is usually a family history. Associated with other autoimmune disease (e.g. thyroid disease) and Vogt–Koyanagi–Harada syndrome (a rare disorder of ocular depigmentation associated with chorioretinal disease, anterior uveitis, ± tinnitus and meningism).

Entropion In-turning of the eye lids due to degenerative changes or secondary to scarring. Most commonly affects the lower lid. ↑ with age (rare <40y). The eyelashes rub on the cornea and irritate the eye. Taping the lower lid to the cheek can give temporary relief. If left untreated, can cause corneal vascularization, ulceration, and infection. Refer for rapid surgical correction.

Ectropion Turning out of the lower eyelid. Causes eye irritation and watering. Most common in the elderly or those with facial nerve palsy (➔ p. 512). Refer for surgery.

Ptosis From the Greek meaning 'to fall', ptosis describes drooping of the upper eyelid. When the normal eye is looking straight forwards, the margin of the upper lid is situated ~2mm above the pupil. Ask the patient to look downwards as far as possible and then upwards as far as possible. The lid margin should move >8mm. The lid margin moves <4mm in patients with severe ptosis. Treat the cause where possible. To determine the cause, look at the pupil:

- Dilated pupil Oculomotor nerve palsy—refer urgently to neurology
- Constricted pupil Horner's syndrome—➔ p. 270
- Normal pupil Old age, congenital, myasthenia gravis, muscular dystrophy, myopathy, botulism

Congenital ptosis Unilateral/bilateral weakness of the levator muscle. Children may compensate by tilting their heads upwards to see better. ~50% have associated superior rectus muscle weakness. Refer for surgical correction if obstructing vision as may cause amblyopia.

Neurological causes of ptosis

- Oculomotor (3rd nerve palsy) If the pupil is dilated, refer urgently to neurology to exclude cerebral haemorrhage or tumour. Ptosis may be partial or complete
- Horner's syndrome ➔ p. 270

Muscular and mechanical causes of ptosis

- **Senile** Most common cause of ptosis—due to age-related changes in the levator muscle. Refer if causing problems
- **Myasthenia gravis** ➔ p. 518
- **Muscular dystrophy** e.g. myotonic or oculopharyngeal dystrophy
- **Myopathy** e.g. Graves' disease
- **Mechanical** Swelling of the eyelid due to allergy or mass effect of tumour

Causes of localized eyelid swelling

- Stye
- Papilloma
- Chalazion
- Xanthelasma
- Sebaceous cyst
- Marginal cyst of Zeis/Moll
- Dermoid cyst—usually upper inner and outer angles of the orbit
- BCC (rodent ulcer)—usually at the lid margin (➔ p. 604)
- Lacrimal gland and lacrimal sac disorders (➔ p. 943)

Stye Common eyelid infection. 2 forms:

External stye (hordeolum externum) Most common form of stye. Infection of a lash follicle or associated gland of Moll (sweat gland) or Zeis (sebum gland) usually by *Staphylococcus aureus*. Confined to the skin and always points outwards. Treat with hot compresses. 💣 Use of antibiotics is controversial. Generally only use topical antibiotic (e.g. chloramphenicol) if associated bacterial conjunctivitis.

Internal stye (hordeolum internum) Abscess of a meibomian gland. Often causes less swelling than external stye. May point inwards onto the conjunctiva (seen as red patch with yellow centre before it bursts) or outwards through the skin. Treat in the same way as external stye with hot compresses ± topical antibiotics as appropriate.

Marginal cyst of Zeis or Moll Non-infected swellings of the glands of Zeis/Moll. No treatment needed, unless troublesome, when refer.

Chalazion/meibomian cyst Following an internal stye, the meibomian gland may become blocked forming a cyst. Cysts may resolve spontaneously but often become infected (treat with hot compresses ± topical/oral antibiotics) and/or chronic. If recurrent infection or chronic cyst refer to ophthalmology for steroid injection or incision/curettage. ❗ If age <7y, refer early; large cysts can affect refraction, causing amblyopia.

Squamous cell papilloma Benign skin tumour—which may form a horn-like lesion. Refer for excision/curettage.

Blepharitis ➔ p. 942 **Xanthelasma** ➔ p. 205

Basal cell carcinoma (rodent ulcer, BCC) ➔ p. 604

Information for patients

Eye Health UK 🖱 www.eyecaretrust.org.uk

Blepharitis and tear duct problems

Blepharitis Chronic, low-grade inflammation of meibomian glands and lid margins. Presents with long history of irritable, burning, dry, red eyes. Eyelids have red margins ± scales on the eyelashes (Figure 25.7). On elevation of the upper lid, look for inflamed meibomian glands. Associated with dry eyes, internal stye (➔ p. 941), and ingrowing eyelashes.

Differential diagnosis Lid papilloma and warts can become inflamed and mimic blepharitis.

Management Prolonged treatment over 2–3mo with regular eye care 3×/wk is needed. Warn patients to persevere as there may be no improvement for up to 2wk:

1. **Warmth** Apply a facial sauna, microwaveable EyeBag®, or hot, moist flannel to the eyes for 5–10min. This is to open the skin pores and meibomian glands. The face should be red after heating.
2. **Massage** Press on the eye lids with a cotton bud to release the meibomian gland secretions—these are seen as thin curly lines. Pressure should not be too light, or firm enough to cause discomfort.
3. **Clean** With the eyes gently closed, use diluted tea-tree oil baby shampoo (10 parts water to 1 part baby shampoo) on a cotton bud to rub along the eyelashes for 15–20s, top and bottom. Clean any remaining shampoo from the lids with a clean face cloth using clear, warm water. Alternatives to baby shampoo include bicarbonate of soda solution or sterile, tea-tree impregnated wipes (Optase®).

After an initial treatment period, it is often necessary to continue to use warm compresses and lid scrubs from time to time to keep the lid scales under control. Treat dry eye symptoms with preservative-free tear supplements, e.g. Liquifilm®.

Exacerbations Treat with topical antibiotics (place a 1cm strip of fusidic acid or chloramphenicol ointment onto a clean finger and rub it into the base of the eyelashes). Oral antibiotics (e.g. doxycycline 50–100mg od) for 3mo may be useful for patients not responding to lid care and topical antibiotics. Topical steroid drops or ointment may sometimes be useful but use only on specialist advice.

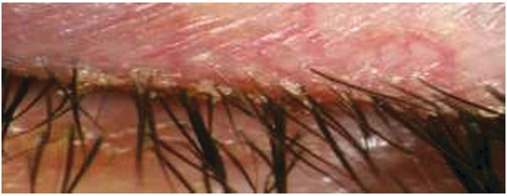


Figure 25.7 Scaling on the eye lashes in blepharitis

Dry eye syndrome (keratoconjunctivitis sicca) Tear secretion ↓ with age. Dry eyes cause eye irritation and redness which is often worse in centrally heated buildings. The eye feels gritty, vision is occasionally blurred, and there is reflex watering of the eye in severe cases. Commonly associated with blepharitis.

Causes ↓ tear production (e.g. age, Sjögren's syndrome); ↑ evaporation of tears (e.g. exposure keratitis).

Management

- Treat with artificial tears, e.g. Viscotears®, Hylo-Care®. If one preparation does not work, try another. Always use preservative-free drops—hypersensitivity can be a problem with prolonged use
- Treat any associated lid disease, e.g. blepharitis
- If simple medication fails, try combined short- and long-acting drops, e.g. Celluvisc® tds and Liquifilm® tds
- Refer to ophthalmology if continuing symptoms despite treatment

❗ Longer-acting drops, (e.g. Celluvisc®, Viscotears®) blur vision for a time. Short-acting drops (e.g. hypromellose 0.3%) only give relief for ~30min and may need very frequent application.

Watering eyes (epiphora) Due to overproduction of tears or outflow obstruction. Caused by corneal irritation (e.g. blepharitis, dry eyes, corneal abrasion, foreign body, conjunctivitis, entropion), iritis, acute glaucoma, ectropion, blocked tear duct.

Acute dacryocystitis Acute infection of the tear sac, can spread to surrounding tissues. Treat immediately with antibiotics, e.g. flucloxacillin. Abscess can form—if it does, surgical drainage is required, so refer.

Chronic dacryocystitis Seen in the middle-aged and elderly. Presents with a watery eye which discharges mucus regularly. The eye does not look inflamed. Refer for syringing of the lacrimal system or surgery.

Infantile dacryocystitis (blocked tear duct) Delay in canalization/obstruction of the lacrimal duct causing persistent watering or sticky eyes in 20% babies. Vision is normal and there is no conjunctival inflammation. If the lower lid conjunctiva is reddened, swab to exclude chlamydia (➔ p. 716).

Management Advise parents to bathe the lids with cooled boiled water. Avoid antibiotic eye drops unless there is clear infection. Spontaneous resolution is the norm. 4% fail to clear by 1y—refer to a paediatric ophthalmologist. Treatment is by probing the duct to clear it.

Information for patients

Eye Health UK 🖱 www.eye-care.org.uk

The red eye and conjunctivitis

⚠ 'Red flag' signs of a potentially dangerous red eye

- ↓ visual acuity
- Pain deep in the eye (*not* surface irritation as with conjunctivitis)
- Absent or sluggish pupil response
- Corneal damage on fluorescein staining
- History of trauma

Refer the patient to be seen by a specialist the same day.

Differential diagnosis Think systematically about the structures within the eye to come to a differential diagnosis—Table 25.2.

Conjunctivitis Inflammation of the conjunctiva is the most common eye problem seen in general practice (Figure 25.8)—1 in 8 children have an episode of acute infective conjunctivitis every year. Presents with unilateral/bilateral red eye with surface irritation; eye discharge (clear, mucoid, or muco-purulent); sticking of the eyelids, especially on waking; no change in visual acuity. Examination may reveal enlarged papillae under the upper eyelid and/or pre-auricular lymph node enlargement.

Bacterial or viral conjunctivitis Clinically difficult to distinguish; doctors get it right only ~50% of the time. Both present with acute red eye—usually starting in one eye and often spreading to involve both, together with watery/purulent discharge. The eyes are often crusted ± stuck together on waking. Visual acuity is not impaired. Both may occur in association with viral URTI.

Management of acute, infective conjunctivitis

- Usually self-limiting condition; 65% settle in 2–5d without treatment; advise patients to bathe the affected eye(s) with boiled, cooled water morning and night, avoid contact lens use, and use simple hygiene measures (e.g. hand washing and not using shared towels)
- If symptoms are not improving in 3–5d, review the diagnosis and consider treatment with topical chloramphenicol qds for 5d. ! Chloramphenicol is available OTC in the UK

⚠ Advise patients to seek medical advice if: ↓ visual acuity, eye becomes painful rather than sore/gritty, significant photophobia, eyelid swelling, or symptoms are not improving in 5d.

Allergic conjunctivitis Bilateral symptoms appear seasonally (e.g. hay fever) or on contact with an allergen (e.g. animal fur). Presents with red, watery, itchy eyes ± photophobia ± family/personal history of atopy. *Signs*: papillae in the lower tarsal conjunctiva and 'cobblestones' under the upper lid.

Management of allergic conjunctivitis Treat with topical or systemic antihistamines (e.g. oral cetirizine, or sodium cromoglicate, nedocromil, or olopatadine eye drops). Avoid topical steroids due to long-term complications (cataract, glaucoma, fungal infection). Consider cold compress and washout with cold water during acute exacerbations. Refer if symptoms are persistent despite treatment, or if vision is affected.

Table 25.2 Differential diagnosis of red eye

Structure	Condition	
Inflammation of the orbit	• Thyroid eye disease/exomphalos (➜ p. 335)	• Tumour (➜ p. 939) • Orbital cellulitis (➜ p. 939)
Lid disease	• Stye (➜ p. 941) • Chalazion (➜ p. 941)	• Blepharitis (➜ p. 942) • Allergic eye disease
Scleral inflammation	• Scleritis/episcleritis (➜ p. 947) • Postoperative inflammation	
Conjunctival disease	• Viral infection • Bacterial infection • Chlamydial infection	• Allergy • Subconjunctival haemorrhage
Corneal disease	• Corneal ulceration (➜ p. 946) • Corneal abrasion (➜ p. 936) • Ophthalmic shingles (➜ p. 947) • Foreign body/trauma (➜ p. 936)	• Arc eye (➜ p. 936) • Dry eye (➜ p. 943)
Uveal/iris inflammation	• Anterior uveitis (➜ p. 947) • Posterior uveitis/toxoplasma	
Other causes of red eye	• Acute glaucoma (➜ p. 955) • Postoperative endophthalmitis (➜ p. 956)	

Ophthalmia neonatorum ➜ p. 716

Herpes simplex keratoconjunctivitis ➜ p. 946–7

Subconjunctival haemorrhage Spontaneous painless localized haemorrhage under the conjunctiva (Figure 25.9). Common in the elderly. Looks alarming but generally painless (may cause some aching of the eye). Clears spontaneously in 1–2wk but may recur. *Associations*: ↑ BP, clotting disorders, leukaemia, ↑ venous pressure. Check BP. If severe/recurrent, check FBC and clotting screen.

❗ Consider referral if follows trauma—especially if the posterior edge of the haemorrhage cannot be seen (may be associated with orbital haematoma, penetrating injury, or orbital fracture).

Pterygium/pinguecula ➜ p. 946

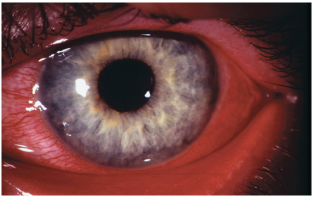


Figure 25.8 Acute infective conjunctivitis

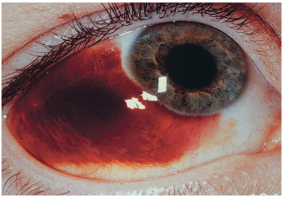


Figure 25.9 Subconjunctival haemorrhage

Corneal, scleral, and uveal disease

Corneal abrasions ➔ p. 936 **Arc eye** ➔ p. 936

Superficial foreign bodies ➔ p. 936 **Corneal arcus** ➔ p. 205

Pterygium Common (Figure 25.10). Found particularly in people who work outdoors in hot, dusty climates. Creamy coloured, raised, triangular plaque on the conjunctiva on either side of the cornea—nasal side > temporal side. No need to treat unless encroaching over the pupil and causing visual loss. If that occurs, refer for surgical excision—recurrence is possible. ❗ Differential diagnosis is carcinoma *in situ*—if any atypical features, refer for excision biopsy.

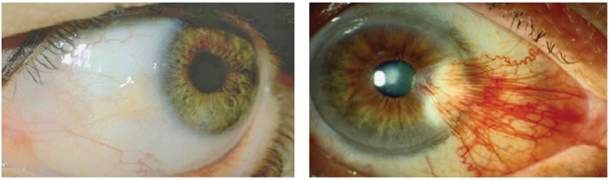


Figure 25.10 Pterygium

Corneal vascularization Growth of blood vessels onto the cornea. Occurs in patients with severe lid disease, rosacea, or due to excessive contact lens wear. If a contact lens wearer, advise to remove contact lenses for at least 2mo. Refer to ophthalmology for specialist management to prevent long-term damage.

Keratitis, keratoconjunctivitis, and corneal ulceration

- Keratitis is inflammation of the cornea
- Keratoconjunctivitis is inflammation of the conjunctiva and cornea

Presentation Presents with a very painful eye, blurred vision, photophobia, and profuse watering. On examination there is ↓ visual acuity, circumcorneal injection (blood vessel dilatation concentrated around the limbus), conjunctivitis (particularly the quadrant most associated with the injury/infection), ± a creamy white, disc-shaped lesion on the central or inferior cornea. The pupil may be small due to reflex miosis. Corneal ulcers stain green with fluorescein—use a bright light with a blue filter to see them.

Causes Bacterial—2° to trauma, foreign body, dry eyes, entropion, blepharitis; viral—herpes simplex, herpes zoster, or adenovirus; fungal; protozoal—history of foreign travel/contact lens wear; non-infective e.g. 2° to autoimmune disease or trauma.

Management Treatment depends on cause. Delay in treatment may result in loss of sight so refer for same-day ophthalmology assessment.

Herpes simplex infection and dendritic ulcer HSV keratitis is common and can be recurrent in the same eye with the virus lying dormant within the trigeminal nerve between attacks. Presents with acute keratitis or keratoconjunctivitis. Occasionally may present as an irritable eye with little discomfort. Examination and fluorescein staining reveals a characteristic corneal ulcer with a delicate branching pattern (dendritic ulcer). Refer for urgent (same-day) ophthalmology opinion. Treatment is with 3% aciclovir ointment 5×/d continued for 3d after healing.

⚠ There is a danger of massive amoebic ulceration and blindness if steroid eye drops are administered to patients with dendritic ulcer.

Ophthalmic shingles Zoster in the ophthalmic branch of the oculomotor (3rd) nerve. Pain, tingling, or numbness around the eye precedes a blistering rash and inflammation. In 50% the eye is affected with conjunctivitis, scleritis, episcleritis, keratitis, iritis, visual loss, and/or oculomotor nerve palsy. Nose tip involvement (*Hutchinson's sign*) makes eye involvement likely (nerve supply is the same as the globe). Prescribe oral aciclovir (800mg 5×/d) and refer immediately. The cornea may become anaesthetic/scarred and require grafting.

Episcleritis The episclera is the thin layer of vascular tissue overlying the sclera. Episcleritis is unilateral in 2 out of 3 cases. It presents with diffuse inflammation of the eye with minimal tenderness and no discharge. Try treatment with an NSAID (e.g. ibuprofen 400mg tds or ketorolac 0.5% eye drops qds). If NSAID is ineffective, refer to ophthalmology for consideration of treatment with steroids.

Scleritis Inflammation of the sclera. Can be unilateral or bilateral. ♀ > ♂. *Peak age*: 40–60y. Affects the anterior or posterior segment and may be diffuse, nodular, or necrotizing. Presents with painful, red eye. Vision may be blurred due to corneal, iris, or posterior segment involvement, and visual acuity ↓. The eye is tender to touch, and may have a deep purple hue. Look for scleral nodules. There may be accompanying uveitis and keratitis.

Associations In ~50%, associated with systemic illness, e.g. herpes zoster, rheumatoid arthritis, systemic lupus erythematosus, polyarteritis nodosum, granulomatosis with polyangiitis, trauma, infection, or surgery.

Management Refer urgently to ophthalmology. Treated with steroids. Complications include cataract, glaucoma, and retinal detachment.

Iritis (anterior uveitis) Most common in young/middle-aged adults. Acute onset of pain, photophobia, blurred vision, and ↓ visual acuity, watering, circumcorneal redness, small or irregular pupil ± keratitic precipitates on the posterior surface of the cornea ± *hypopyon* (anterior chamber pus, causing a white 'fluid-level' line). Pain ↑ as eyes converge and pupils constrict. May be secondary to corneal graft rejection or eye infections, e.g. toxoplasmosis, herpes virus keratitis. In 30% associated with seronegative arthropathies, e.g. ankylosing spondylitis.

Management Refer urgently to ophthalmology. Complications include posterior synechiae (irregular pupil shape), glaucoma, and cataract. Relapses are common.

Visual field loss and blindness

Visual field loss Not all patients who have a visual field loss are aware of it. ! If you suspect visual field loss, refer to a community optometrist or ophthalmology for formal field testing. *Causes*: Figure 25.12.

Blindness is defined as inability to perform any work for which eyesight is essential (not the total absence of sight). In practice this means <3/60 vision (may be >3/60 if patient has severe visual field defect, e.g. glaucoma). 157,000 people are registered blind in England.

Partial sightedness Does not have a standard definition but usually implies vision in the range 3/60—6/60; 155,000 people are registered partially sighted in England.

Major causes of blindness in the UK

- Elderly Macular degeneration; glaucoma
- Younger patients Diabetic retinopathy; uveitis; inherited retinal disease; retinovascular disease

Registration of blindness and partial sight Voluntary in the UK. Refer patients for low-vision assessment. Application is made by a consultant ophthalmologist to social services. The registration process for children in Scotland is slightly different.

Support Many patients benefit from links with national support organizations that provide information, and active local organizations who support the blind and partially sighted with drivers and guides.

↓ visual acuity and driving ➔ p. 98. Different rules may apply for specific field defects—seek specialist advice.

Colour blindness Congenital colour blindness—Figure 25.11.

Impaired colour recognition Occurs later in life. Red is the most common colour affected. May be an early sign of an optic nerve disorder. Patients complain of colour looking 'washed out' (desaturated) in one eye compared to the other—refer.

Prohibits certain employment (e.g. airline pilot). Inherited as a sex-linked characteristic. ♂:♀ ≈ 20:1. The Ishihara test is a series of 24 cards of coloured dots comprising a number against a contrasting background. Dots are paired to detect patterns of colour blindness. Loss of red/green discrimination is most common.

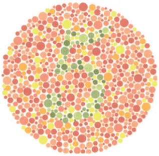


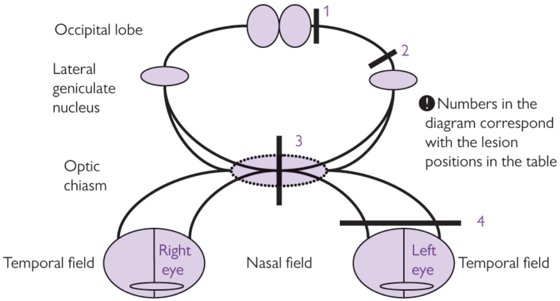
Figure 25.11 Congenital colour blindness

Information and support for patients and carers

LOOK (for families of blind/visually impaired children) ☎ 01432 376314 🖱 www.look-uk.org

Partially Sighted Society ☎ 01302 965195 🖱 www.partsight.org.uk

Royal National Institute for the Blind (RNIB) Information and talking book service. ☎ 0303 123 9999 🖱 www.rnib.org.uk



❗Numbers in the diagram correspond with the lesion positions in the table

Defect (lesion position)	Description and causes
Cortical blindness	Normal eyes; normal papillary responses; no conscious vision. *Cause:* Bilateral damage to the visual cortex - usually CVA
Homonymous hemianopia (1)	Half the visual field is affected symmetrically in both eyes. Macular fibres may be preserved (macular sparing) if the posterior cerebral artery is functional. *Cause:* Strokes involving the middle cerebral artery
Quadrantinopia (2)	Loss of a homonymous (symmetrical) quadrant of vision indicates temporal lobe disease with superior defect and parietal lobe disease with inferior loss. *Causes:* Vascular events, tumours, trauma
Bitemporal hemianopia (3)	The temporal side of the visual field is affected in both eyes. If one nerve is completely affected, a junctional scotoma results. *Causes:* Compressive chiasmal lesions e.g. pituitary tumour, craniopharyngioma, or meningioma
Altitudinal defect (4)	Field defect respecting the horizontal. *Cause:* Optic nerve disease e.g. optic neuropathy, optic neuritis
Enlarged blind spot (4)	Blind spot is enlarged if the optic disc is enlarged. *Causes:* Papilloedema, disc inflammation, infiltration with lymphoma
Central scotoma (4)	Loss of central vision with normal visual field around it. May be unilateral or bilateral • Bilateral causes: toxic (e.g. tobacco), B_{12} deficiency, MS, age related macular degeneration, inherited • Unilateral causes: glioma of optic nerve, vascular lesion
Tunnel vision	Loss of peripheral vision in all directions. *Causes:* Glaucoma, retinitis pigmentosa, retinal detachment, functional visual loss (visual fields having no anatomical correspondence)
Loss of vision from one eye	Due to lesions of the retina or optic nerve anterior to the optic chiasm. *Causes:* Retinal detachment, retinal vein occlusion, optic neuropathy, infiltration of the nerve, demyelination, compression of the nerve

Figure 25.12 Visual field loss, position of lesion, and causes

Sudden loss of vision in one eye

⚠ Always refer as an emergency to ophthalmology—unless you are certain loss of vision is migraine or results from a stroke/TIA.

Causes of sudden loss of vision covered elsewhere

- Acute glaucoma (➲ p. 955)
- Stroke/amaurosis fugax (➲ p. 534)
- Migraine (➲ p. 528)
- Temporal arteritis (➲ p. 498)
- Wet AMD (rapid rather than sudden loss of vision—➲ p. 952)

Retinal vein occlusion Incidence ↑ with age. More common than arterial occlusion. *Presents with*:

- Sudden loss of vision in one eye—typically on waking (branch retinal vein occlusion causes partial visual loss) ± afferent pupil defect
- Fundus like 'a stormy sunset'—scattered haemorrhages, engorged veins, disc swelling ± cotton wool spots
- ~90d after retinal vein occlusion, the eye may become painful due to neovascular glaucoma

Causes

- Glaucoma
- Arteriosclerosis
- ↑ BP
- Polycythaemia
- Hypercholesterolaemia
- ↑ homocysteine

Management Refer as an emergency to ophthalmology. Laser treatment and/or intraocular anti-vascular endothelial growth factor (anti-VEGF) medication may prevent neovascular glaucoma, and vitreous haemorrhage due to retinal neovascularization. Macular oedema may be helped by intraocular steroids or intraocular anti-VEGF injections.

Retinal artery occlusion Usually due to thromboembolism. Sudden visual loss in one eye (counting fingers or light perception) and afferent pupil defect. The retina appears white ± cherry red spot at the macula. A retinal embolus may be visible. Exclude temporal arteritis (➲ p. 498).

⚠ *If the patient presents <1h after onset* Applying then releasing firm eyeball pressure can sometimes dislodge an embolus into one of the smaller branches and thus preserve some vision.

Management Refer as an emergency to ophthalmology. There is no reliable treatment. Optic atrophy and blindness is the usual outcome. Treat any risk factors for atherosclerosis or embolism, i.e. ↑ BP, hyperlipidaemia, smoking, DM, carotid/cardiac disease.

Vitreous haemorrhage Presents with sudden ↓ in vision, loss of red reflex, and difficulty visualizing the retina. *Risk factors*: DM with new vessel formation, bleeding disorders, retinal tear/detachment, central retinal vein occlusion, trauma, head injury, tumour. Refer urgently to ophthalmology. Treatment is with vitrectomy and repair of retinal damage with laser- or cryotherapy.

Retinal detachment Affects 1 in 7000 people each year. *Presentation*:

- Painless loss of vision—'like a curtain' coming across the vision
- Rate of detachment can vary. Upper retinal detachments tend to occur more quickly—causing loss of lower part of vision

- 50% have premonitory symptoms—flashing lights or floaters before eyes due to abnormal retinal stimulation prior to the detachment
- If the macula is detached, central vision is lost and may not completely recover—even after retinal reattachment
- Examination reveals visual field loss (± central visual loss), afferent pupil defect, and a grey retina which may balloon forwards

Causes

- Idiopathic
- Trauma
- DM
- After cataract surgery
- Myopia
- Retinopathy of prematurity
- Inherited eye disease

Management Refer urgently for treatment to secure the retina.

Floaters Small dark spots in the visual field usually caused by opacities in the vitreous. Floaters continue to move when the eye comes to rest. *Risk factors*: myopia, cataract operation, trauma. Usually harmless and may settle with time but patients may benefit from vitrectomy if floaters are interfering with vision (e.g. preventing reading).

> ⚠ Sudden showers of floaters in one eye ± flashing lights can indicate retinal detachment which may be difficult to see on examination. Floaters associated with eye pain/inflammation may indicate posterior uveitis.

General rules for referral

- If longstanding floaters/flashes then no need for referral
- If symptoms are of recent onset (<6wk) and no other symptoms, refer urgently to ophthalmology outpatients
- If symptoms are of recent onset (<6wk) and associated with any visual field loss, ↓ acuity, or pain/inflammation of the eye, refer as an ophthalmology emergency

Optic neuritis Disc swelling due to inflammation or demyelination. Presents with rapid visual loss (hours–days) and ↓ colour vision (red desaturation); discomfort on eye movements; temporary worsening of symptoms when hot; optic disc swelling. Refer urgently to ophthalmology for confirmation of diagnosis. Steroids may help in severe cases. Visual loss usually stabilizes after week 2 and recovers over 6wk.

Causes Multiple sclerosis (1 in 4 patients with MS present with optic neuritis—➔ p. 540); DM; viral infections, e.g. influenza, measles, chickenpox; familial, e.g. Leber's disease.

Anterior ischaemic optic neuropathy (ANION) Occurs when the short ciliary arteries are damaged. 2 forms:

- Arteritic Due to arterial inflammation (e.g. temporal arteritis, SLE)
- Non-arteritic Results from arterial emboli

Presentation Central vision drops suddenly and irreversibly. Examination reveals a complete or altitudinal visual field defect. The disc appears swollen and pale ± haemorrhages. May be accompanied by symptoms of the underlying condition (e.g. temporal arteritis—➔ p. 498).

Management Refer as an ophthalmology emergency. Treatment depends on cause.

Gradual loss of vision

Causes of gradual loss of vision covered elsewhere

- Chronic glaucoma (➔ p. 954)
- Cataract (➔ p. 956)
- Diabetic retinopathy (➔ p. 328)

Age-related macular degeneration (AMD) Most common cause of blindness in the UK—2% of people aged >65y are blind in one or both eyes due to AMD. Always a bilateral disease but one eye is usually more severely affected than the other. *Risk factors*: ↑ age, +ve family history, smoking, ↑ BP, AMD in the other eye, poor diet, lack of exercise.

Presentation Difficult to detect in primary care. Signs are often minimal. Use Snellen chart, near-vision test, and an Amsler grid (Figure 25.13) to test vision. *Symptoms*:

- In all cases, there is deterioration/distortion of central vision—affects reading/face recognition first—worse with changes in lighting
- A dark patch that rapidly fades may be noticed on waking—can be interpreted as 'seeing a shadowy figure' and be very frightening
- With severe visual loss patients may see visual hallucinations—usually of faces or stars. These can also be very frightening

Dry (geographic) AMD All patients start with this form of AMD. Caused by atrophy of the neuroretina. The cells of the macula break down resulting in drusen formation (yellowish lipid deposits). As number/size of drusen ↑, central vision ↓.

Wet AMD Accounts for 50% of blindness due to AMD. In some patients with dry AMD, drusen lift the retinal pigment epithelium away from its blood supply. New blood vessels grow from the choroid and may bleed forming scars → irreversible loss of central vision.

Management If progressive loss of vision, refer to ophthalmology for confirmation of diagnosis—urgently if recent onset or rapid ↓ in vision. For those with dry AMD and loss of vision, treatment with AREDS2 food supplement (containing vitamin C, vitamin E, zinc, copper, lutein, and zeaxanthin) may ↓ progression[R]. Treatment of other coexisting conditions (e.g. cataract and glaucoma) can also help. Provision of visual aids, registration of blindness, and social support are also important.

Vascular endothelial growth factor inhibitors e.g. aflibercept, ranibizumab. Effective treatment for patients with active wet AMD and visual symptoms. Administered in 2° care by intravitreal injection (directly into the eye) every 4–12wk depending on preparation used and treatment response.

With usual glasses on (if needed), hold the grid at normal reading distance

Cover one eye and focus at the dot in the centre of the grid. Repeat with the other eye

If the lines appear to be wavy, distorted or broken in any way, suspect macular degeneration.

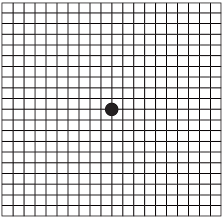


Figure 25.13 How to use an Amsler grid

Central serous retinopathy (CSR) Typically occurs in hypermetropic middle-aged patients. Vision is blurred/distorted particularly for reading. Caused by serous leakage of fluid from abnormal choroidal vessels. Generally self-limiting, but can be bilateral and chronic. Refer.

Macular hole Typically ♀ in mid 60s. Presents with gradual central visual loss/distortion and colour loss. If in the non-dominant eye, may be relatively asymptomatic. Refer to ophthalmology. Vision takes 4–12mo to recover following surgical treatment.

Macular dystrophies Several inherited retinal diseases (e.g. Best's disease; Stargardt's macular dystrophy; Bull's eye maculopathy) present with progressive loss of central vision either in early adulthood or aged 40–60y. Ask if there is a history of visual loss in a family.

Retinitis pigmentosa Familial disorder resulting in retinal degeneration. Usually noticed in adolescence and progresses to blindness. 2 forms—autosomal dominant form is more common and milder than the autosomal recessive form. Presents with night blindness, loss of visual field, difficulty in light adaptation, and gradual loss of central vision. On examination there are black pigment flecks in the retina, optic atrophy, and attenuated blood vessels. There is no effective treatment.

Epiretinal membrane May present with distortion and blurred central vision, particularly for near vision. Associated with peripheral vascular disease, retinal detachment, branch retinal vein occlusion, uveitis, trauma, or tumour. Refer to ophthalmology for vitrectomy and membrane peel.

Optic atrophy *Signs*: gradual visual loss; pale optic disc. *Causes*: glaucoma, MS, ischaemia (e.g. retinal artery occlusion), retinal damage (choroiditis, retinitis pigmentosa), toxic (tobacco amblyopia, methanol, arsenic, quinine). Refer to ophthalmology or neurology to confirm diagnosis.

Compressive lesions of the optic pathway e.g. meningioma, glioma, abscess, AV malformation can cause visual field defects; type depends on the site of the lesion—➜ p. 949.

Retinoblastoma ➜ p. 884

Retinal melanoma Most common tumour affecting the eye. Usually detected during routine examination by an optometrist. Other presentations include gradual central visual loss and/or retinal detachment. Can affect the iris, ciliary body, or choroids. Refer for urgent ophthalmology opinion.

Retinal metastases Can occur from tumours of the breast, lung, or kidney. Appear as pale elevations of the choroid. Symptoms include visual ↓ and retinal detachment. Refer urgently if suspected.

Further information

AREDS trial 🖱 www.areds2.org
NICE (2018) Age-related macular degeneration. 🖱 www.nice.org.uk/guidance/ng82

Information and support for patients

Macular Disease Society ☎ 0300 3030 111 🖱 www.macularsociety.org
RP Fighting Blindness ☎ 0845 123 2354 🖱 www.rpfightingblindness.org.uk

Glaucoma

Chronic simple glaucoma (open-angle) (COAG) Common. Affects ~2% of all >40y olds. Accounts for ~1 in 4 ophthalmology outpatient appointments and 10% of new blindness registrations.

Risk factors

- ↑ intraocular pressure (IOP) >21mmHg—the major risk factor—but 30% of newly diagnosed glaucoma patients have 'normal' pressure
- Family history (↑ risk ×10)
- ↑ age
- Black race
- Abnormal BP (↑ in elderly)
- Myopia
- ↑ plasma viscosity

❗ Steroid use (systemic or topical in or close to the eye) can cause ↑ IOP.

Presentation May be detected during routine optometrist examination or through routine screening for diabetes or for patients with family history. Otherwise patients present late as glaucoma is asymptomatic and visual acuity is preserved until visual fields are severely impaired. *Signs*: optic nerve damage (glaucomatous disc cupping), visual field loss (sausage-shaped blind spots), and ↑ IOP.

Variants

- Ocular hypertension ↑ IOP with no field loss
- Normal tension glaucoma Field loss, disc cupping, but normal IOP

Management Advise all patients >40y to have regular optometry check-ups. Those with a family history of glaucoma should have biannual checks of their IOPs (*tonometry*), and annual visual field checks, at an optician from 40y of age. Refer patients with ↑ pressures, or in whom you notice (or are doubtful about) disc cupping to ophthalmology for assessment. Patients with ↑ IOP must be followed up lifelong. Aim is to ↓ intraocular pressure to slow disease progression—even with 'normal' pressures.

Medical treatment ❗ If prescribing >1 drop, adherence and tolerance may be better with a combination drop:

- Topical prostaglandin analogue (e.g. latanoprost od in the evening)—↑ outflow of aqueous. First-line treatment for those with IOP ≥24 mmHg at risk of visual impairment within their lifetime[N]
- Topical β-blocker (e.g. timolol 0.25% bd)—↓ aqueous secretion. Caution in patients with asthma or heart failure. Often combined with a topical prostaglandin analogue. *Side effects*: allergy and dry eyes
- Topical carbonic anhydrase inhibitor (e.g. dorzolamide tds, or bd if in combination with a β-blocker)—↓ aqueous secretion. *Side effects*: blurred vision, tiredness, dyspepsia
- Topical α-agonist (e.g. brimonidine bd)—↓ aqueous secretion and ↑ outflow. *Side effects*: local reactions, headache, dry mouth, tiredness

Surgery Considered when the 'target' IOP is not met with medical treatment (especially in patients <50y).

Acute closed-angle glaucoma (ACAG) Uncommon, Affects 0.1% of patients >40y. Typically ♀, elderly, and long-sighted with early cataract. Closed-angle glaucoma may present in 1 of 3 ways:

- **Latent** Usually picked up when screening the opposite eye after an episode of acute/subacute glaucoma. The patient is asymptomatic and IOP normal, but the anterior chamber is shallow with a narrow angle
- **Subacute** Episodic haloes around bright lights, impaired vision, ± frontal headache/eye pain. Attacks are precipitated by the pupil dilating, e.g. at night or entering a darkened room and relieved by sleep or entering a brighter environment. Examination between attacks is normal but during an attack, the pupil is semi-dilated and cornea slightly clouded. Patients with subacute glaucoma are at risk of an acute attack
- **Acute** Blockage of aqueous drainage from the anterior chamber causes a sudden ↑ in IOP from 15–20 to 60–70mmHg. There may be a history of previous subacute attacks. The patient complains of eye pain with acute loss of vision in 1 eye ± abdominal pain/nausea/vomiting

Examination Vision ↓; cornea looks hazy (due to oedema); pupil is fixed and dilated (often slightly oval in shape with long axis vertical); circumcorneal redness; eyeball feels hard (due to ↑ pressure); poor fundal view ± cataract.

Management Refer acute or subacute glaucoma as an emergency to ophthalmology. Specialist treatment is with meiosis to open drainage channels (e.g. pilocarpine 4% drops) and acetazolamide ± apraclonidine and/or latanoprost drops to ↓ aqueous production. Surgery or laser treatment (peripheral iridotomy) to allow free aqueous circulation is undertaken once intraocular pressure has been ↓. Patients may need prophylactic surgery on the contralateral eye to prevent ACAG in that eye too. ACAG may damage the trabecular meshwork and patients are at risk of developing chronic glaucoma following an attack. Regular check-ups are necessary.

Neovascular or secondary glaucoma May occur in patients with diabetic retinopathy, central or branch retinal vein obstruction, or ocular ischaemia. Blood vessels grow across the iris and the iridiocorneal angle, preventing fluid drainage. Pressures can be very high (40–70mmHg) and the patient may suffer pain from corneal oedema. Treatment is surgical and, in severe cases, if the eye is blind, it is removed.

Congenital glaucoma 1 in 10,000 live births. ♂ > ♀. Usually bilateral. Presents with irritation of the eye (watering, rubbing), photophobia, large eyes with large, fixed pupils ± cloudy cornea. Refer urgently for paediatric ophthalmic opinion. Surgery is needed to prevent blindness.

Further information

NICE (2017) Glaucoma: diagnosis and management. 🖱 www.nice.org.uk/guidance/ng81

Information and support for patients

International Glaucoma Association ☎ 01233 648170 🖱 www.glaucoma-association.com

Cataract

Lens opacity is found in 75% of >65y olds. Most do not need treatment. Cataract is lens opacity associated with visual loss.

Risk factors for cataract

- Old age
- DM
- +ve family history
- Prolonged steroid treatment
- ↑ BP
- Excessive alcohol
- Smoking
- Prenatal rubella/toxoplasma (congenital cataract)
- Hypocalcaemia
- Eye trauma
- Radiation exposure

Presentation

- Blurred vision and gradual loss of vision
- Dazzles and halos around objects—especially in sunlight
- Frequent spectacle changes due to changing refractive index

❗ Unilateral cataract may not be noticed by the patient—but loss of binocular vision affects judgement of distance.

Signs A shadow in the red reflex/absent red reflex; difficulty visualizing the fundus.

Types of adult cataract There are 3 main types of adult cataract:

- **Nuclear cataract** Tends to present with ↓ contrast and ↓ colour intensity. Patients may have difficulty recognizing faces but reading is often well preserved (Figure 25.14)
- **Cortical cataract** May not cause any symptoms initially. With time may cause difficulty reading, light scatter, and problems with glare when driving. Most disabling in low light when the pupil is dilated
- **Posterior subcapsular cataract** Tends to affect younger patients. Causes problems reading and driving in good light—less disabling when light is poor and the pupil is dilated

Primary care management of adult cataract Check fasting blood glucose to exclude DM. Advise patients to have their visual acuity checked regularly. Refer to ophthalmology for consideration of surgery if:

- ↓ sight—or any other symptom resulting from cataract, *and*
- Impairment of lifestyle, social functioning, driving, or independence, *and*
- Willingness to have surgery, if appropriate

Cataract surgery >90% of cataract extraction procedures are carried out in people ≥60y. Removal of the natural lens ± posterior chamber lens implantation. Usually a day case procedure under LA. Healing takes 2–6wk depending on the technique used. 75–95% without other ocular pathology have 6/12 vision or better 3mo post-op. Patients need testing for new spectacles 6wk post-op.

Early complications of cataract surgery

- **Intraocular infection (endophthalmitis)** Rare (0.1%)—presents with pain and blurred vision ± red eye ± tenderness. Refer back to the operating surgeon urgently— antibiotics injected within 2–3h can preserve vision. Delayed referral (>12h) will lead to blindness
- **Intraocular bleeding**
- **Damage to other eye structures** Posterior capsule rupture (1–3%); trauma to the iris; wound gape/iris prolapse

- Broken or protruding sutures Cause sensation of a foreign body on the cornea, or pain—may need to be removed

Late complications of cataract surgery

- Posterior capsule opacification Results in cloudy vision. Affects up to 40% of patients who have had cataract surgery—usually <2y post-op. Treatment is with laser therapy to create a hole in the capsule
- Dysphotopsia Most common cause of post-op dissatisfaction (2.5–12%). Intraocular light artefacts resulting from reflection of light from the intraocular lens. May result from lens edge effects or be due to the anatomy of the eye
- Retinal detachment 1 in 150 operations—more likely if high myopia; can occur weeks or even years post-op
- Glaucoma Closed or open angle—➔ p. 954
- Rare Cystoid macular oedema, bullous keratopathy, uveitis

Cataract in children May be:

- Congenital Genetic, metabolic (e.g. galactosaemia), or result from *in utero* infection (TORCH—toxoplasmosis, rubella, cytomegalovirus, herpes simplex)
- Developmental Genetic (e.g. Down's syndrome), metabolic (e.g. galactokinase deficiency)
- Acquired Trauma, radiotherapy, metabolic (e.g. DM)

Presents with squint, white pupil, nystagmus, amblyopia, or loss of binocular vision. If detected, refer urgently to ophthalmology for specialist management.

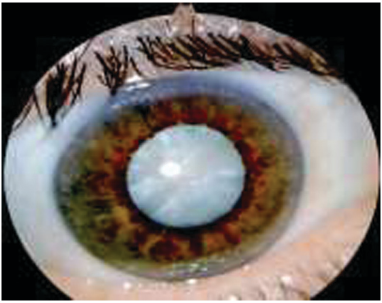


Figure 25.14 Mature nuclear cataract

Further information

NICE (2017) Cataracts in adults: management. 🖱 www.nice.org.uk/guidance/ng77

Information for patients

Eye Health UK 🖱 www.eye-care.org.uk
Royal College of Ophthalmologists 🖱 www.rcophth.ac.uk

Refraction errors and squint

Glasses check Look through the patient's glasses:

- If image is magnified (prescription '+')—the patient is long-sighted
- If image is reduced (prescription '−')—the patient is short-sighted

Amblyopia (lazy eye) Poor vision in the absence of ocular or visual pathway disorder. Squint, ptosis, cataract, unequal refractive errors, or astigmatism can cause the image from one eye to be disregarded. If this persists >7–8y of age, it becomes irreversible. Treatment is with glasses ± patching, and squint surgery if necessary.

Refraction errors

Hypermetropia (long sight) Most common refractive error. Common in infants and lessens with age. Distant objects focus behind the retina. Ciliary muscle contraction (to make the lens more convex) is needed to focus the image. This can lead to convergent squint, eye tiredness, and headache. Convex lenses are used for correction.

Myopia (short sight) Distant objects focus in front of the retina. There is often a +ve FH. Concave lenses are used to correct the defect. Contact lenses may be necessary in high myopia (>−8 dioptres). Myopia is unusual <6y old and tends to worsen until the late teens. Regular (6-monthly) eye checks are needed to ensure correct lenses are prescribed. In adults, ↑ myopia can indicate developing cataracts. High myopia (8–20 dioptres) predisposes to retinal detachment—these patients should have an annual eye examination (more frequent if floaters).

Astigmatism Curvature across the cornea or lens differs in the vertical/horizontal planes. Objects are distorted longitudinally or vertically. Lenses can be used to correct this defect.

Presbyopia Age-related loss of accommodation. The lens becomes less easy to deform from 45–65y. Focusing on close objects (accommodation) is more difficult and glasses may be needed for near work (e.g. reading).

Refractive procedures Increasingly being undertaken as an alternative to spectacles. *LASIK* (laser assisted *in situ* keratomileusis) is a combination of surgery and laser therapy. It can be used for higher degrees of refractive error and astigmatism. Complications are rare. Refractive lens exchange is increasingly popular in those <50y old.

Non-paralytic squint Abnormality of coordinated eye movement; 3% of children have a congenital squint. Due to an imbalance in the muscles of the eye; there is full range of eye movement in both eyes and no double vision. Note the light reflexes from different parts of the cornea—they should be symmetrical. If not, there is a squint. Squint may be convergent (esotropia) or divergent (exotropia). Esotropia (Figure 25.15) is most common and often associated with long-sightedness.

Predisposing factors FH of squint, high refractive errors, neurological disease (e.g. cerebral palsy), cataract, Down's/Turner's syndrome, retinoblastoma, optic atrophy, craniofacial anomalies, retinal disease.

Childhood screening for squint ➔ p. 836

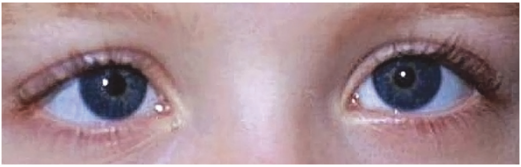


Figure 25.15 Esotropic squint

Management Refer to ophthalmology. Without treatment children with squint risk developing amblyopia, failure of binocular vision, and long-term visual problems. Visual maturity occurs at 7–8y. Eye patching, correction of refractive errors (spectacles), and realignment surgery can improve sight up to this age.

Paralytic squint Caused by damage to the extraocular muscles or the nerves supplying them. Usually acquired and caused by cranial nerve palsy. Results in diplopia—maximal when looking in the direction requiring the action of the paralysed muscle. Image from the eye that is not moving correctly is peripheral to the normal-eye image. Refer for urgent neurology/ophthalmology opinion (same-day review if ill with headache). If sinister causes have been excluded, management involves treatment of the underlying condition ± patching and/or prism spectacles ± surgery.

Pseudosquint

- **Wide epicanthic folds** Give the appearance of a squint—corneal reflections are symmetrical
- **Intermittent deviation of the eyes in neonates** Common. Check red reflex is present. Normally settles by 3mo—squint after this time is significant. Refer

Gaze palsy Inability to perform coordinated movements of the 2 eyes together in the same direction. In all cases, refer to neurology—treatment depends on cause (Table 25.3).

- Horizontal gaze palsy—loss of conjugate eye movements to one side
- Vertical gaze palsy—loss of conjugate eye movements upwards

Table 25.3 Ocular nerve palsies

Nerve	Effect of paralysis	Causes of nerve palsy
3rd Oculomotor nerve	Ptosis and ophthalmoplegia—eye looks down and out Surgical causes are also associated with pain, proptosis, and pupil dilation	*Surgical*: berry aneurysm (posterior communicating artery); cavernous sinus lesions *Medical*: microvascular disease, e.g. DM
4th Trochlear nerve	Superior oblique muscle is paralysed. Causes diplopia and torticollis. The eye cannot look down and inwards	Trauma (30%), DM (30%), idiopathic
6th Abducens nerve	Lateral rectus paralysed. Causes diplopia. The eye is turned in and cannot move laterally from the midline	Tumour, trauma to the base of the skull, vascular

Drugs for the eye and contact lenses

Drugs and the eye Many eye complaints can be treated with topical medication. Ointments last longer in the eye than drops but can cause blurring of vision—they are particularly useful at night.

❗ Intolerance/allergy to preservatives in eye drops is common and can result in sore, red eyes. If suspected, preservative-free unit dose vials may be an alternative.

Mydriatics (e.g. tropicamide, cyclopentolate) Dilate the pupil and cause cycloplegia thus causing blurred vision. They are used to dilate the pupil for examination and to prevent adhesions to the lens in iritis. They can precipitate acute closed-angle glaucoma in susceptible patients.

Miotics (e.g. pilocarpine) Constrict the pupil and ↑ aqueous drainage. They are used in glaucoma. They can cause systemic side effects, e.g. sweating, ↑ BP, pulmonary oedema.

Local anaesthetic drops (e.g. proxymetacaine) Can help examination of painful eyes and foreign body removal. Protect the eye with an eye pad until the anaesthetic has worn off to prevent corneal damage (corneal reflex is suppressed).

Steroid eye drops Used in scleritis, episcleritis, iritis. Prescribe only after slit lamp examination and on the advice of an ophthalmologist. Can cause severe eye damage if used when a dendritic ulcer is present. Long-term use may cause glaucoma, thinning of the cornea/sclera, and may facilitate fungal infection.

β-blocking drops (e.g. timolol, betaxolol) Used in glaucoma. Beware of systemic side effects—bronchospasm, bradycardia.

α2 receptor agonists (e.g. brimonidine) Used in glaucoma. May cause dry mouth, headache, fatigue.

Prostaglandin analogues (e.g. latanoprost, bimatoprost) Used in glaucoma. May cause lash growth and ocular inflammation.

Lubricants (e.g. Hylo-Forte®, Viscotears®, Liquifilm®) Can be prescribed without preservative as unit dose vials. Helpful for dry eyes.

Antibiotics (e.g. chloramphenicol, ofloxacin, fusidic acid) For topical treatment of infection. Antibiotics should generally be given as drops, enabling clearance through the naso-lacrimal system. In severe infections 2-hourly drops should be used, reducing to qds after 48h. Antibiotic preparations (e.g. chloramphenicol, fusidic acid) can become contaminated with bacteria so should be changed regularly, Chloramphenicol can be prescribed 'preservative-free' if needed.

Mast cell stabilizers (e.g. sodium cromoglicate 2% qds, nedocromil 2% bd) Useful for hayfever and other allergic conjunctivitis.

Contact lenses 20% are worn because they are more suitable for the eye condition than spectacles, 80% for cosmetic/convenience reasons. Some are worn just to change eye colour. Contact lenses are used in high myopia or hypermetropia, presbyopia, and after cataract removal because thick spectacle lenses cause visual field distortion. They are also useful when the cornea has been damaged, e.g. after ulceration or trauma and in keratoconus (a rare corneal degenerative disease).

Types of lens Hard, gas-permeable (larger hard lenses are designed to allow air to reach the cornea) and soft lenses are available. Some soft lenses are 'daily disposable' or 'monthly disposable'. Hard and gas-permeable lenses can correct for minor astigmatism, normal soft lenses cannot as the lens is too flexible. A high astigmatism requires spectacles or a special (toric) soft lens (delicate and needs careful cleaning). Patients with poor tear secretion do not tolerate contact lenses well.

Care of lenses Careful cleaning of the lenses and contact lens container is vital—particular solutions are used for each type of lens and these should not be interchanged. Contact lenses can be stained by fluorescein or rifampicin—ask before prescribing.

Complications

- Eye infection
- Corneal abrasion or vascularization (painful, watery eye after lens removal)
- Sensitization to cleaning agents (redness, stinging, swollen eyelids)
- Giant papillary conjunctivitis
- Losing the lens within the eye
- Keratitis
- Acanthamoeba infection

Chapter 26

Mental health

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Mental health assessment

Assessing a patient with mental health problems in primary care can be challenging. Aims (may need >1 appointment to achieve):

- Establish a constructive relationship to enable effective two-way communication and serve as the basis for subsequent encounters
- Assess the patient's emotions/attitudes and risk to self and/or others
- Determine if the patient has a mental disorder and, if so, which
- Find out (where possible) what caused the mental disorder
- Explore how best it might be treated

Psychiatric history

Informants GPs may be approached by concerned relatives/friends. Talk to them but maintain confidentiality, establish circumstances/concerns, relevant past history, and the patient's pre-morbid personality.

The consultation Try to review old notes *before* seeing the patient. Use open questions at the start, becoming directive when necessary—clarify, reflect, facilitate, listen. Be open and ready to ask about suicide, sex, drugs, etc. Ask about:

- **Presenting complaint** Chronological account, past history of similar symptoms. Ask directly about thoughts of suicide and self-harm
- **Family history** Psychiatric illness (depression or mania, psychotic disorder), recent loss/serious illness of a family member, bereavement, suicide or attempted suicide, alcoholism, drug use, dementia
- **Personal history** Abuse (emotional, physical, sexual), domestic violence, substance misuse, serious illness (including past psychiatric and major physical illness), recent significant events (e.g. life stressors, childbirth)
- **Attitudes and beliefs** How does the patient see him/herself? What does the patient think is wrong? How does the patient think other people view the situation? What does the patient want you to do?
- **Occupation** Employment status, happy in job, access to suicide means, e.g. chemicals, drugs, firearms
- **Home situation** Housing, relationships (including whether an informal carer), social support, finances etc.

Mental state examination

Check:

- **Appearance and behaviour** Signs of self-neglect or malnutrition, eye contact, rapport, movements, agitation, or aggression
- **Speech** Spontaneity, volume, tone, rate, amount, continuity (flight of ideas, loosening of associations)
- **Mood** Depressed or elevated. Consider screening for depression, e.g. screening questions for patients with chronic disease (➔ p. 173), screening questions for pregnant (➔ p. 787) and postnatal women (➔ p. 819), PHQ-9 (➔ p. 980)
- **Anxiety** Generalized or situation specific. Consider screening for anxiety, e.g. GAD2 (➔ p. 971)
- **Thinking** Form, content, flow, possession
- **Perception** Illusions, hallucinations, pseudohallucinations
- **Cognition** Cognitive screen, e.g. 6 CIT (➔ p. 989)
- **Insight** Patient's understanding of his/her illness, its effects, and need for treatment

Action

- Summarize the history back to the patient and give an opportunity for the patient to fill in any gaps or clarify any points
- Draw up a problem list and management plan with the patient
- Set a review date
- Challenge the stigma of mental illness where necessary; remember 1 in 4 people experience mental health problems each year

⚠ High-risk groups for psychiatric illness

Women

- More vulnerable to depression and eating disorders
- During pregnancy and in the postpartum period
- When looking after children <5y old, especially lone parents who also go out to work
- When subjected to domestic violence
- During the menopause

Men More at risk of completed suicide ➔ p. 1100.

People with long-term physical health problems e.g.

- Diabetes
- Heart disease
- Chronic disabling lung disease
- Cancers—especially when newly diagnosed
- Dementia or learning difficulty—may be difficult to assess
- Disabling neurological disorders: stroke, PD, MS, MND

Substance misusers Drug misuse; alcohol dependence.

People in contact with the criminal justice system

Veterans of the Armed Services Particularly PTSD (➔ p. 976).

People suffering adverse life events

- Bereavement
- Relationship break-up
- Unemployment
- Financial problems

Minority ethnic groups More likely to suffer mental health problems due to social and economic deprivation, isolation from their usual culture, racism, and past exposure to war or torture (>50% of refugees have mental disorders); less likely to seek help.

Carers All carers are at risk of depression (~40% of carers of stroke victims are depressed). This starts early after the onset of caregiving. It is good practice to:

- Identify all carers and mark their records
- Check carers' mental and physical health annually
- Inform carers that they are entitled to a needs assessment
- Ask patients if you can share information with their carers
- Inform carers about support groups and carer centres

Residents of residential care homes 50% have depression; often overlooked due to co-morbidities.

Mental health symptoms and signs

Acute confusion (delirium) ➲ p. 988 **Anxiety** ➲ p. 970

Depression ➲ p. 978

Abnormal beliefs Decide whether a belief is normal in the context of the patient. If not, decide if the belief is a:

- Delusion i.e. a. belief that does not seem to have a rational basis and which is not amenable to argument, *or*
- Overvalued idea i.e. belief that preoccupies the sufferer but is not of delusional intensity or obsessional in nature

Compulsions Forced behaviours repeated despite inappropriateness, or unreasonableness, and associated discomfort in response to an obsession. Can be disabling, e.g. repeated handwashing hundreds of times a day. Obsessive–compulsive disorder—➲ p. 974.

Abnormal perceptions *Consider*:

- Illusions Misinterpretation of information, e.g. seeing a coat on a hanger and interpreting it as a person. Can happen if ↓ level of consciousness or occasionally if visual impairment, particularly AMD
- Hallucinations See following section
- Pseudohallucinations Vivid perception which is recognized as not being real, e.g. delirium tremens, Charles Bonnet syndrome in AMD
- Depersonalization Feeling of being unreal—like an actor playing yourself. Associated with a wide range of mental illness (e.g. depression, schizophrenia) and drugs
- Derealization Feeling of everything around you being unreal—like in a dream. Often coexists with depersonalization

Hallucinations Sensory experiences in the absence of external stimuli. May be visual, auditory, gustatory, olfactory, or tactile.

- Visual/tactile/auditory hallucinations suggest mental illness:
  - Visual and tactile hallucinations suggest organic disorder, e.g. dementia, acute delirium, metabolic encephalopathy, drug abuse
  - Auditory hallucinations suggest psychosis
- Hallucinations experienced when the patient is falling asleep (hypnagogic hallucination) or waking up (hypnapompic hallucination) are features of narcolepsy
- Olfactory and gustatory hallucinations Often occur together. May be suggestive of psychosis but also occur with temporal lobe epilepsy and olfactory bulb tumours

Thought disorders Consider disorders of:

Content

- Ideas of reference Patients may feel they are being watched by everyone around them/stand out from the crowd; media content, e.g. television or radio, refers to the patient, or that others are talking/thinking about them. Becomes a delusion of reference when insight is lost. Associated with schizophrenia, but also severe mood disorders, and acute and chronic cognitive impairment
- Delusional beliefs ➲ p. 967

Flow

- **Flight of ideas** Leaps from idea to idea. There is always some association between ideas but may seem odd, e.g. rhymes. Easily distractible. Associated with manic illness
- **Perseveration** Typically a persistence of a verbal response beyond what is apparently intended, expected, or needed. Usually organic, e.g. associated with dementia, brain damage, CVA
- **Loosening of association** Series of thoughts appear only distantly (or loosely) related to one another or completely unrelated. Associated with schizophrenia
- **Thought blocking** Abrupt and complete interruption in the stream of thought leaving a blank mind. Can occur mid-sentence. Involuntary. Associated with schizophrenia

Form

- **Preoccupation** The patient thinks about a topic frequently but can terminate the thoughts voluntarily. Common symptom, e.g. in anxiety states. Ask about preoccupation with suicide in depressed patients
- **Obsession** Thought or image repeated in spite of its inappropriateness or intrusiveness and associated discomfort. The thought and efforts to stop it ↑ anxiety and can be disabling

Passive thought control Strongly associated with schizophrenia:

- **Thought insertion** Thoughts do not belong to the patient but have been planted there by someone else. One of the first-rank symptoms of schizophrenia
- **Thought withdrawal** Opposite of thought insertion. The patient perceives a thought is missing and has been removed by someone else. A first-rank symptom of schizophrenia
- **Thought broadcasting** The patient believes his/her thoughts can be heard by other people—either directly or via the newspapers, radio, etc. Associated with schizophrenia

Delusions Beliefs held unshakably despite available counter-evidence and which are inexplicable in view of circumstances and background. The belief is usually (but not always) false in basis.

- **Primary delusions** Belief arrives in the head fully formed, e.g. thought insertion; strongly suggestive of schizophrenia
- **Secondary delusions** Belief arises on the basis of experience, e.g. someone who has lost their job several times through no fault of their own may believe they are unemployable; a delusion of inappropriate guilt is another common example

Paranoid delusions Delusions (usually primary) which concern the relationship between the patient and other people. Associated with schizophrenia, depressive states, and cognitive impairment.

- **Delusions of persecution** Most common type of paranoid delusion. Belief that a person or organization is intentionally harassing or inflicting harm upon the patient. Associated with schizophrenia, depressive states, and cognitive impairment
- **Delusions of grandeur** Beliefs of possessing exaggerated power, importance, knowledge, or ability. Associated with mania/schizophrenia

Psychological therapies

In the UK, the 'Improving Access to Psychological Therapies' (IAPT) programme in England and similar programmes in other areas of the UK have dramatically ↑ access to psychological therapies. These may be used alone or in combination with drug therapies. Referral criteria vary.

Who should be referred? Consider referral for patients with:

- Stress
- Grief reactions
- Eating disorders
- Body dysmorphic disorder
- Depression
- Phobias
- Somatization
- Obsessive–compulsive disorder
- Anxiety
- Panic disorder
- Personality disorders
- Post-traumatic stress disorder
- Medically unexplained symptoms

Problem-solving therapy (PST) Can be an effective tool for use in the GP surgery. Involves drawing up a list of problems, and generating and agreeing solutions, broken down into steps, for patients to work on as homework between sessions. Patients re-attend for review of success and to consider alternative approaches if goals are not met—➔ p. 981.

Cognitive behaviour therapy (CBT)

- **Behavioural therapies** Aim to change behaviour. Usually the therapist uses a system of graded exposure (systematic desensitization) combined with teaching a method of anxiety reduction
- **Cognitive therapy** Focuses on people's thoughts and the reasoning behind their assumptions on the basis that incorrect assumptions (that are often unconscious) → abnormal reactions which then reinforce these assumptions further (a vicious cycle)

The patient learns to recognize negative or unhelpful thinking patterns. By enabling the patient to be more aware of this, and teaching ways to challenge cognitive errors, more helpful thinking styles can result. Patients must then practise re-evaluating their thoughts, and associated behaviours. At least as effective as drug treatment[N].

Individual non-facilitated self-help

Computerized CBT (CCBT) Useful for patients with mild symptoms or who cannot attend face-to-face sessions:

- Free online interactive CCBT, e.g. Living Life to the Full (🖱 www.livinglifetothefull.com)
- Other CCBT, e.g. Mood Gym (🖱 https://moodgym.com.au); Beating the Blues (🖱 www.beatingthelues.co.uk); Silvercloud (🖱 www.silvercloudhealth.com). Local access arrangements vary; a fee may be payable or subscriptions purchased for patients by local PCOs

Bibliotherapy For patients who are not computer literate, self-help books based on CBT techniques are an alternative, e.g.:

- Gilbert P (2009). *Overcoming depression: A Self-Help Guide Using Cognitive Behavioural Techniques*. ISBN: 9781849010665
- Kennerley H (2014). *Overcoming Anxiety: A Self-Help Guide Using Cognitive Behavioural Techniques*. ISBN: 9781849018784

Guided self-help Based on a CBT approach and available through psychological therapy services. Helpful for patients with mild symptoms. Uses books/printed/online materials under the supervision of a trained facilitator who introduces, monitors, then reviews the outcome of each treatment. Usually there is minimal contact with the facilitator (<3h).

Mindfulness-based cognitive therapy Skills training programme designed to prevent the recurrence of depression. Aims to bring a focus on the moment, e.g. by focusing on breathing or a visual image, and empty the mind of negative thoughts. ↓ relapse by >50% in the first year after treatment. Aimed at patients who have ≥3 depression relapses.

Behavioural activation Therapist and patient work together with the aim of identifying effects that the patient's behaviour might have on symptoms, mood, and problems. Problematic behaviours are then addressed. Techniques may include reducing avoidance, activity scheduling, graded exposure, and initiating positively reinforced behaviours.

Interpersonal therapy (IPT) Individual or group therapy concentrating on the difficulties that arise in maintaining relationships with others. Focuses on current, not past, relationships, and works on the premise that if conflicts are resolved, both relationships and mood will improve. Useful for patients who can identify relationship difficulties.

Psychoeducational groups Group therapy can be used to explore depression or chronic physical health conditions, e.g. diabetes. Run by trained practitioners, they also involve the element of peer support.

Applied relaxation Group or individual therapy that establishes the cues that herald feelings of anxiety and helps the patient to relax when those cues are felt. Effective treatment for anxiety states.

Counselling Usually reflective listening encouraging patients to think about and try to resolve their own difficulties. Little evidence of benefit or cost-effectiveness^{N} but if there is a specific, identifiable cause for the patient's symptoms, counselling directed at the cause may be helpful, e.g.:

- Relationship breakdown—counselling is available through organizations such as RELATE (🖱 www.relate.org.uk)
- Bereavement—counselling is available via organizations such as CRUSE (☎ 0808 808 1677; 🖱 www.cruse.org.uk)
- Debt—counselling is available from Citizens Advice, National Debtline (☎ 0808 808 4000), or Step Change Debt Charity website (🖱 www.stepchange.org)

Further information

NICE (2009, updated 2018) Depression in adults: recognition and management. 🖱 www.nice.org.uk/guidance/cg90

NICE (2011) Common mental health problems: identification and pathways to care. 🖱 www.nice.org.uk/guidance/cg123

NICE (2011, updated 2019) Generalised anxiety disorder and panic disorder in adults. 🖱 www.nice.org.uk/guidance/cg113

SIGN (2010) Non-pharmaceutical management of depression in adults: A national clinical guideline. 🖱 https://www.sign.ac.uk/sign-114-non-pharmaceutical-management-of-depression.html

Anxiety

Anxiety is only considered abnormal when it occurs in the absence of a stressful trigger; impairs physical, occupational, or social functioning; and/or is excessively severe or prolonged. Features include:

Psychological symptoms:

- Fearful anticipation
- Irritability
- Sensitivity to noise
- Restlessness
- Poor concentration
- Worrying thoughts
- Insomnia and/or nightmares
- Depression
- Obsessions
- Depersonalization
- Fear of losing control/dying

Physical symptoms:

- Dry mouth
- Tremor
- Dizziness
- Epigastric discomfort
- Difficulty swallowing
- Frequent/loose motions/flatulence
- Chest discomfort/constriction
- Difficulty breathing/hyperventilation
- Palpitations/awareness of missed beats
- Frequency/urgency of micturition
- Sexual dysfunction
- Menstrual problems
- Paraesthesiae
- Tinnitus
- Headache

Generalized anxiety disorder (GAD) Long-term condition, fluctuating in severity and nature, often beginning in adolescence. Lifetime prevalence ~5%. Key features are excessive, difficult-to-control worry about a number of events/activities occurring on most days for ≥6mo. Consider if history of anxiety/worry, or if frequent attender with chronic physical health problem, or no physical health problem but needing reassurance about somatic symptoms, or repeated worrying about a wide range of different issues.

Associations

- Anxiety is often accompanied by depression (➔ p. 978) and may be a feature of early schizophrenia
- Other conditions which can cause and/or mimic symptoms of anxiety include drug/alcohol use or withdrawal, caffeine abuse, thyrotoxicosis, hypoglycaemia, temporal lobe epilepsy, phaeochromocytoma (rare)

Assessment Check TFTs. Use GAD-2 score (Box 26.1) for screening:

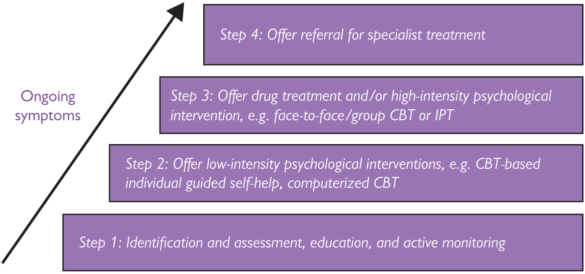
- If score ≥3, consider anxiety disorder
- If score <3 but you suspect anxiety is a factor, ask '*Do you find yourself avoiding places or activities and does this cause you problems?*'—if the answer is yes, consider anxiety disorder

Management

- Avoid caffeine, excess alcohol, and illicit drugs
- Use a stepped treatment approach (Figure 26.1)[N]
- Provide information about self-help organizations/support groups
- Try to identify causes of anxiety
- Consider using an anxiety scale (e.g. Hamilton Anxiety Scale) to record baseline morbidity and progress at follow-up

Psychological therapies ➔ p. 968

- Low intensity—individual non-facilitated self-help; individual guided self-help; psychoeducational groups
- High intensity—CBT or applied relaxation


Step 4: Offer referral for specialist treatment
Step 3: Offer drug treatment and/or high-intensity psychological intervention, e.g. face-to-face/group CBT or IPT
Step 2: Offer low-intensity psychological interventions, e.g. CBT-based individual guided self-help, computerized CBT
Step 1: Identification and assessment, education, and active monitoring
Ongoing symptoms

❗ Offer the least obtrusive, most effective treatment first

Figure 26.1 Stepwise approach to GAD management

Drug treatment

- SSRIs (e.g. sertraline 50–200mg od; escitalopram 10–30mg od)—warn patients medication may take >1wk to work and of possible side effects (short-term ↑ in anxiety; GI symptoms). If >60y or other risk factors for GI bleeding, consider co-prescribing a PPI
- Follow up every 2–4wk in the first 3mo then every 3mo
- If drug treatment is effective, continue for >1y (↓ relapse rates)
- If no benefit, consider alternative SSRI/SNRI or adding a psychological therapy; pregabalin is an option if unable to tolerate SSRI/SNRI. Do not offer antipsychotic medication
- Avoid benzodiazepines except for acute crises; restrict use to <4wk

⚠ Patients <30y may have ↑ suicidal thoughts when they start an SSRI/SNRI—warn about this risk and follow up <1wk after starting medication and then weekly for 1mo to monitor suicide/self-harm risk. Benzodiazepines may be useful for this first 1–2wk period.

Refer to specialist mental health services If severe anxiety with marked functional impairment plus:

- Risk of self-harm/suicide, or
- Significant co-morbidity (e.g. substance misuse, personality disorder, or complex physical health problems), or
- Self-neglect, or
- Inadequate response to step 3 interventions

Box 26.1 GAD-2 short screening tool

Over the last 2wk, how often have you been bothered by any of the following problems?	Not at all	Several days	> half the days	Nearly every day
1. Feeling nervous, anxious, or on edge	0	1	2	3
2. Not being able to stop or control worrying	0	1	2	3

GAD-2 was developed by Spitzer RL, Williams BW, Kroenke K et al, with an educational grant from Pfizer Inc.

Panic disorder Panic attacks are very common, but panic disorder is uncommon: lifetime prevalence—1% of ♂; 3% of ♀. *Associations*: depression (56%); GAD; agoraphobia; substance abuse; suicide (↑ risk).

- **Panic attack** Period of intense fear with characteristic symptoms. Can be spontaneous or situational
- **Panic disorder** Chronic disorder; recurrent panic attacks associated with persistent fear of having (or the consequences of) another attack

Symptoms Anxiety builds up quickly and unexpectedly without a recognizable trigger and patients often describe an intense feeling of apprehension or impending disaster. Common associated symptoms:

- Shortness of breath/smothering sensations
- Choking
- Palpitations and ↑ heart rate
- Chest discomfort or pain
- Sweating
- Dizziness, unsteadiness
- Nausea or abdominal pain
- Depersonalization/derealization
- Numbness or tingling sensations
- Flushes or chills
- Trembling or shaking
- Fear of dying
- Fear of doing something crazy

Examination Obvious distress; sweating; tachycardia; hyperventilation. ↑ BP is common and usually settles. Otherwise examination is normal.

Differential diagnosis Alcohol withdrawal; drug misuse/withdrawal; other psychiatric disorders (e.g. psychosis); hyperthyroidism; temporal lobe epilepsy; cardiac arrhythmia; hypoglycaemia; hyperparathyroidism; phaeochromocytoma (very rare).

Management of acute panic attack ➔ p. 1081

Management of panic disorder Use a stepped treatment approach:

- **Step 1** Recognition and diagnosis. Educate about the condition, signpost to support in the community, and discuss treatment options. Commence active monitoring. Avoid alcohol, illicit drugs, and caffeine
- **Step 2** Treatment in primary care—offer (in order of effectiveness) psychological therapy (CBT), drug treatment, or self-help (bibliotherapy or CCBT)—choice depends on severity of symptoms, co-morbidities, and patient preference
- **Step 3** Consideration of alternative treatment—if one step 2 treatment is ineffective, change to or add another
- **Step 4** Offer referral for specialist treatment if ≥2 primary care treatments have failed

Drug treatment ❗ Do not use benzodiazepines for treatment of patients with panic disorder—associated with poorer long-term outcome.

- Offer SSRI, e.g. paroxetine, citalopram, or SNRI, e.g. venlafaxine. Warn about possible transient ↑ in anxiety on starting treatment. Minimize initial side effects by starting at low dose and ↑ slowly. Review in <2wk and at 4, 6, and 12wk
- If SSRI/SNRI is not suitable or ineffective, offer a TCA (e.g. imipramine, clomipramine) or another form of treatment
- If effective, continue for ≥6mo, reviewing every 8–12wk
- Minimize discontinuation symptoms by tapering dose over time

Phobias As GAD but limited to certain situations. 2 main features:

- **Avoidance** Of the circumstances that provoke anxiety
- **Anticipatory anxiety** If there is a prospect of meeting that situation

Simple phobia Inappropriate anxiety in the presence of ≥1 object/situation, e.g. flying, enclosed spaces, spiders. Common in early life; most adult phobias are a continuation of childhood phobias. *Lifetime prevalence*: 4% ♂; 13% ♀.

Management Only treat if symptoms are frequent, intrusive, or prevent necessary activities. Exposure therapy is effective. Obtain through psychological therapy services or the private sector, e.g. British Airways 'Fear of flying' course. Do not use benzodiazepines.

Social phobia (social anxiety disorder) Intense/persistent fear of being scrutinized or negatively evaluated by others leads to fear and avoidance of social situations (e.g. using a telephone, speaking in front of a group). Significantly disabling; not just shyness. May be generalized (person fears most social situations) or specific (related to certain activities only). Ask about safety-seeking behaviour. *Management*:

- **Drug therapy** SSRIs—continue ≥12mo, or long term if symptoms remain unresolved, there is a co-morbid condition (e.g. depression, GAD, panic attacks), a history of relapse, or early onset
- **Psychological therapies** CBT (cognitive restructuring) ± exposure

Agoraphobia Peak age of onset is 20–40y with an initial panic attack. Subsequently, panic attacks, fear of fainting, and/or loss of control are experienced in crowds, away from home, or in situations from which escape is difficult. Avoidance results in patients remaining within their homes where they feel safe. Other symptoms include depression, depersonalization, and obsessional thoughts.

Management Difficult to manage in general practice. Diagnosis may be delayed as patients will not visit the surgery; ongoing management is complicated by refusal to be referred for specialist care. Prognosis is best when there is good marital/social support. *Options*:

- **Behaviour therapy** e.g. exposure, coping with panic attacks. Home visits may be required, but should be resisted as part of therapy
- **Drug treatment** SSRIs (citalopram and paroxetine are licensed); TCAs (e.g. imipramine, clomipramine); MAOIs. Relapse rate is high. Benzodiazepines may be used sparingly if frequent panic attacks, particularly if initiating other treatment, but beware of dependence

Further information

NICE (2011) Common mental health problems: identification and pathways to care. 🖱 www.nice.org.uk/guidance/cg123

NICE (2011, updated 2019) Generalised anxiety disorder and panic disorder in adults. 🖱 www.nice.org.uk/guidance/cg113

NICE (2013) Social anxiety disorder: recognition, assessment and treatment. 🖱 www.nice.org.uk/guidance/cg159

Patient information and support

Anxiety Care 🖱 www.anxietycare.org.uk
No More Panic 🖱 www.nomorepanic.co.uk
Triumph over phobia (TOP) UK 🖱 www.topuk.org

Other anxiety-type disorders

Stress ➲ p. 976 **Post-traumatic stress disorder** ➲ p. 976

Mixed anxiety and depression Combinations of anxiety and depression are common—particularly among women. Prevalence ~10%. When anxiety and depression occur together symptoms are more severe, there is ↑ functional impairment, illness is more chronic/persistent, and there is poorer response to treatment. Treat as for anxiety and/or depression depending on the predominating features. Refer for psychiatric assessment if management strategies are not working.

Obsessive–compulsive disorder (OCD) Recurrent obsessive thoughts and compulsive acts. Lifetime prevalence ~2% although minor obsessional symptoms are more common. ♂:♀ ≈2:3. Tends to present in young adults. Patients may have symptoms for years before seeking help as they know that their thoughts/actions are irrational and are embarrassed to tell anyone. Relatives may highlight the problem.

Features

- Obsessional thinking Recurrent persistent thoughts, impulses, and images causing anxiety or distress
- Compulsive behaviour Repetitive behaviours, rituals, or mental acts done to prevent or ↓ anxiety
- Other features Indecisiveness and inability to take action, anxiety, depression, and depersonalization

Screening questions[N]

- Do you wash or clean a lot?
- Do you check things a lot?
- Is there any thought that keeps bothering you that you would like to get rid of but can't?
- Do your daily activities take a long time to finish?
- Are you concerned about orderliness or symmetry?
- Do these problems trouble you?

Management in adults

- Mild functional impairment Offer short CBT (<10h) including exposure–response prevention (ERP) or group therapy
- Moderate functional impairment Offer more intensive CBT (>10h) or drug therapy (SSRI, e.g. fluoxetine 20–40mg od)
- Severe functional impairment Offer psychological therapy + drug treatment. If inadequate response at 12wk, offer a different SSRI or clomipramine. Refer if symptoms persist. Other TCAs and SNRIs should not be used

Management in children and young people

- Mild functional impairment Consider guided self-help. Include support and help for family and carers
- Moderate/severe functional impairment Offer CBT including ERP adapted to patient's age in a group or individual setting. Refer if symptoms do not improve. Drug therapy should only be initiated in secondary care

Body dysmorphic disorder (BDD) Preoccupation with an imagined defect in appearance or markedly excessive concern over a slight physical anomaly. *Prevalence*: 0.5–0.7%.

Features Time-consuming behaviours, e.g. mirror gazing, comparing self to others, camouflage, reassurance seeking, and skin picking.

Screening questions[N]

- Do you worry about the way you look and wish you could think about it less?
- What specific concerns do you have about your appearance?
- On a typical day, how many hours a day is it on your mind? (>1h is excessive)
- What effect does it have on your life?
- Does it make it hard to do your work or be with friends?

Management As for OCD, however, all children/young people should be offered CBT.

Somatization Physical symptoms in response to emotional distress. Characterized by an excessive preoccupation with bodily sensations combined with a fear of physical illness. Common feature of depression, anxiety, schizophrenia, and substance use.

Medically unexplained symptoms ➔ p. 178

Somatization disorder Chronic condition. History of numerous unsubstantiated physical complaints. Starts at <30y and often persists many years. ♀:♂ ≈10:1; lifetime prevalence 0.1–0.2% although mild symptoms are much more common. Presents with:

- >2y history of multiple symptoms with no physical explanation
- Refusal to be reassured that there is no explanation for the symptoms
- Impaired social/family functioning due to these symptoms and/or associated behaviours

Management of somatization disorder

- Reattribution involves acknowledging/taking symptoms seriously, offering necessary examination and investigations, asking about psychosocial problems, and explaining the link between symptoms and stress
- Treat co-morbid psychiatric problems (e.g. depression, anxiety, panic)
- Beware of risks of drug interaction—self-medication with multiple OTC (or even prescription) drugs is common
- Beware of side effects of medication—these patients do not tolerate prescribed drugs well and have a heightened awareness of side effects
- Refer to the specialist mental health team if risk of suicide, marked functional impairment, impulsive or antisocial behaviour

Further information

NICE (2005) Obsessive–compulsive disorder and body dysmorphic disorder: treatment. 🖱 www.nice.org.uk/guidance/cg31

Patient information and support

OCD Action ☎ 0845 390 6232 🖱 www.ocdaction.org.uk

Royal College of Psychiatrists Health advice leaflets. 🖱 https://www.rcpsych.ac.uk/healthadvice/atozindex.aspx

- Obsessive–compulsive disorder
- Obsessive–compulsive disorder in children and young people

Chronic stress

We all suffer from stress and, most of the time, the pressures of everyday life are a motivating force. A problem only arises when those pressures exceed the individual's ability to cope with them.

Causes of stress Virtually anything we do can cause stress. The most common causes of stress-related morbidity in the UK are:

- Work problems
- Family problems
- Financial problems
- Legal problems
- Exam stress

The stress epidemic >11 million working days are lost each year in the UK due to stress (45% of all sickness absence). The Health and Safety Executive estimates ~0.5 million people in the UK are experiencing work-related stress at a level they believe is making them ill; up to 5 million people feel 'very' or 'extremely' stressed by their work; and work-related stress costs society >£5 billion every year. Occupational stress is most likely to affect those working in the health, social work, and education sectors.

Presentation Most patients do not consult their GP with stress unless they feel it is affecting their health. Common symptoms include:

- Mood swings
- Anxiety
- Depression
- Low self esteem
- Dry mouth
- Fatigue and/or lethargy
- Poor or ↑ appetite
- ↑ smoking, alcohol, and/or caffeine consumption
- Sleep disturbance
- Headaches
- Loss of libido
- Menstrual abnormalities
- Poor concentration and/or memory
- Other unexplained aches/pains, e.g. muscular pains, chest pains
- Worsening of pre-existing conditions, e.g. irritable bowel syndrome, eczema, asthma, psoriasis, migraine

The GP's role Is to identify stress as a cause of presenting symptoms; educate patients about stress and links between symptoms and stress; identify sources of stress; provide support and self-management strategies (➔ p. 968); treat medical problems arising out of stress, e.g. depression; and provide certification if stress is so great that unable to work.

Post-traumatic stress disorder (PTSD) May occur in 25–30% of those who have experienced/witnessed traumatic events, e.g. major accident, fire, assault, military combat. It can affect people of all ages.

Symptoms Most develop symptoms immediately after the event but it is common for sufferers not to present until months/years afterwards. In ~15%, onset of symptoms is delayed. 65% experience chronic symptoms:

- Intrusive recollections Thoughts; nightmares; flashbacks
- Avoidant behaviour Of people, places, situations, or circumstances resembling/associated with the event; refusal to talk/think about the event; excessive rumination about questions about the event (e.g. Why me? How could it have been prevented?)
- Arousal ↑ anxiety/irritability, insomnia, ↓ concentration, ↑ vigilance
- Numbing of emotions Inability to experience feelings; feelings of detachment; giving up previous activities; amnesia for parts of the event

Associations Depression, anxiety; drug/alcohol abuse and dependence.

Management Treat any other associated psychiatric illness.

- **Watchful waiting** If mild symptoms have been present <4wk. Be supportive and listen. Arrange follow-up in <1mo
- **Trauma-focused psychological treatment** CBT and/or eye movement desensitization and reprocessing (EMDR). Refer if severe symptoms <4wk, or ongoing intrusive symptoms >4wk after trauma
- **Drug treatment** e.g. paroxetine and sertraline are licensed. Not first-line treatment. Reserve for those refusing psychological therapy, or with continuing symptoms despite psychological therapy

❗ Single-session debriefing interventions after traumatic events are unhelpful. Do not offer non-trauma-focused therapy.

The stressed GP ➔ p. 22

10 tips for chronic stress relief

- Ensure you get enough sleep and rest; avoid using sleeping tablets to achieve this—➔ p. 168
- Look after yourself and your own health, e.g. don't skip meals, sit down to eat, take time out to spend time with family and friends, make time for hobbies and relaxation, do not ignore health worries
- Avoid using nicotine, alcohol, or caffeine as a means of stress relief
- Work off stress with physical exercise—↓ levels of adrenaline released and ↑ release of natural endorphins which lead to a sense of well-being and enhance sleep
- Try relaxation techniques
- Avoid interpersonal conflicts—try to agree more and be more tolerant
- Learn to accept what you can't change
- Learn to say 'no'
- Manage your time better—prioritize and delegate (see following section); create time buffers to deal with unexpected overruns and emergencies
- Try to sort out the cause of the stress, e.g. talk to line manager at work, arrange marriage or debt counselling, arrange more child care

Time management made easy This technique aims to transform an overwhelming volume of work into a series of manageable tasks:

- Make a list of all the things you need to do
- List them in order of genuine importance
- Note whether you really need to do the task, what you need to do personally, and what can be delegated to others
- Note a timescale in which each task needs to be done, e.g. immediately, within a day, within a week, month, etc.

Further information

Health and Safety Executive (HSE) 🖱 www.hse.gov.uk/stress

NICE (2018) Post-traumatic stress disorder. 🖱 www.nice.org.uk/guidance/ng116

Patient advice and support

International Stress Management Association (UK) 🖱 www.isma.org.uk

Stress Management Society ☎ 0203 142 8650 🖱 www.stress.org.uk

Depression

2.3 million people suffer from depression in UK at any time. ♂:♀ ≈1:2.

Recognition ~30–50% cases are not detected, although most are mild cases, more likely to resolve spontaneously. Challenge any stigma of mental illness.

Screening questions for depression^N

- During the last month, have you often been bothered by feeling down, depressed, or hopeless?
- During the last month, have you often been bothered by having little interest or pleasure in doing things?

If +ve response to either question, investigate further, e.g. with PHQ-9 (Figure 26.2).

Causes and co-morbidity Associated with:

- **Psychiatric disorders** e.g. anxiety, alcohol abuse, substance abuse, eating disorders
- **Physical disorders** e.g. PD, MS, dementia, thyroid disorders, Addison's disease, hypercalcaemia, RA, SLE, cancer, HIV and other chronic infections, cardio- and cerebrovascular disease, learning disability
- **Drugs causing symptoms of depression** β-blockers, anticonvulsants, Ca^{2+} channel blockers, corticosteroids, oral contraceptives, antipsychotic drugs, drugs used for PD (e.g. levodopa)

History

- Onset including precipitating events
- Nature of symptoms, severity, and effect on life
- Past history of similar symptoms, mood elevation (may be bipolar disorder) or other mental health problems; previous response to treatment
- Current life events—stressors at home and at work
- Family and social history including quality of interpersonal relationships, living conditions, and social isolation
- Coexistent medical conditions
- Current medication—prescribed and non-prescribed

! Sleep disturbance and fatigue have high predictive value for depression and should prompt enquiry about other symptoms.

Symptoms Present >50% of the time in the past 2wk. 2 key features:

- Depressed mood *and/or*
- ↓ interest or pleasure, which must be disabling to the patient

Other symptoms

- Change in appetite/weight
- Insomnia or hypersomnia
- Fatigue or loss of energy
- Poor concentration
- Poor appetite or overeating
- Low energy or fatigue
- Low self-esteem
- Psychomotor agitation/retardation
- Sense of worthlessness or guilt
- Recurrent thoughts of death/suicide
- Feelings of hopelessness
- Poor concentration or difficulty making decisions

Seasonal affective disorder ➲ p. 994

Name: Date:

Over the last 2 weeks, how often have you been bothered by any of the following problems? (use ✓ to indicate your answer)	Not at all	Several days	More than half the days	Nearly every day
1. Little interest or pleasure in doing things	0	1	2	3
2. Feeling down, depressed, or hopeless	0	1	2	3
3. Trouble falling or staying asleep, or sleeping too much	0	1	2	3
4. Feeling tired or having little energy	0	1	2	3
5. Poor appetite or overeating	0	1	2	3
6. Feeling bad about yourself—or that you are a failure or have let yourself or your family down	0	1	2	3
7. Trouble concentrating on things, such as reading the newspaper or watching television	0	1	2	3
8. Moving or speaking so slowly that other people could have noticed. Or the opposite—being so fidgety or restless that you have been moving around a lot more than usual	0	1	2	3
9. Thoughts that you would be better off dead, or of hurting yourself in some way	0	1	2	3
Add columns:				
		Total:		
10. If you ticked off *any* problems, how *difficult* have these problems made it for you to do your work, take care of things at home, or get along with other people?		Not difficult at all Somewhat difficult Very difficult Extremely difficult		

Figure 26.2 The Patient Health Questionnaire (PHQ-9)

Patient Health Questionnaire (PHQ-9) by Kroenke K, Spitzer RL, Williams JB. from *J Gen Intern Med*. 2001 Sep;**16**(9):606–13. Development funded by Pfizer Ltd.

Examination

- **General appearance** Self-neglect, smell of alcohol, weight ↓
- **Assessment of mood** Looks depressed and/or tired, speech monotone or monosyllabic, avoids eye contact, tearful, anxious or jumpy/fidgety, feeling of distance, poor concentration, etc.
- **Psychotic symptoms** Hallucinations, delusions, etc. ➔ p. 966

Assessing severity of depression Can be done using a depression symptom count or patient self-complete measure, such as the PHQ-9.

- **Subthreshold depression** <5 symptoms of depression (PHQ-9 score <5)
- **Mild depression** ≥5 symptoms of depression that result in only mild functional impairment (PHQ-9 score of 5–9)
- **Moderate depression** symptoms or functional impairment are between ‘mild’ and ‘severe’ (PHQ-9 score of 10–14 indicates moderate depression; PHQ-9 score of 15–19 indicates moderately severe depression)
- **Severe depression** most symptoms and the symptoms markedly interfere with functioning ± psychotic symptoms (PHQ-9 score ≥20)

⚠ **Assessment of suicidal intent** ❗ Always ask patients directly about suicidal ideas and intent (➔ p. 1100). *Risk factors for suicide*:

- ♂ > ♀
- Age 40–60y
- Living alone
- Divorced > widowed > single > married
- Unemployment
- Chronic physical illness
- Past psychiatric history
- Recent admission to psychiatric hospital
- History of suicide attempt/self-harm
- Alcohol/drug misuse
- Family history of suicide

Cultural considerations Some cultures have no terms for depression. Patients may present with physical symptoms (somatization) or use less familiar ‘cultural specific’ terms to describe depressive symptoms, e.g. ‘sorrow in my heart’.

Management of depression[N] Use a stepped care approach starting at the step most appropriate for the patient.

Step 1 All patients—assess severity. Provide education about depression and information about support available to both the patient and carers/family members as appropriate. Discuss treatment options.

If sleep is a problem Discuss sleep hygiene (Box 7.2, ➔ p. 169): establish regular sleep/wake times; avoid excess eating, smoking, or alcohol before sleep; create a proper environment for sleep; take regular exercise.

Subthreshold or mild depression Active monitoring—watchful waiting for <2wk to see if spontaneous recovery occurs. Simple problem-solving strategies may be useful in the GP surgery (Figure 26.3).

Step 2 Persistent subthreshold or mild/moderate depression.

- **Low-intensity psychosocial interventions** (➔ p. 968) Individual guided self-help; CCBT; structured group exercise programme (e.g. exercise prescription)
- **Group-based peer support** For people with a shared chronic physical health problem
- **Drug treatment** Do not use routinely for subthreshold/mild depression—consider if: past history of moderate/severe depression; subthreshold symptoms that have been present >2y; subthreshold/mild depression that persists after other interventions

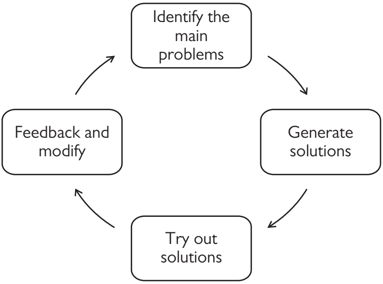


Figure 26.3 Simple problem-solving strategy to use in the surgery

Step 3 Moderate/severe depression or mild/subthreshold depression that has not responded to treatment.

- **High-intensity psychological interventions** (➔ p. 968) Individual or group-based CBT, interpersonal therapy, behavioural activation, behavioural couples therapy (if relationship difficulties)
- **Drug treatment** Usually SSRI as first-line agent
- **Combined treatments** Antidepressant medication + high-intensity psychological intervention (CBT or interpersonal therapy)
- **Collaborative care** For people with physical health problems. Case management supervised by a mental health professional providing education + psychological/pharmacological treatments with follow-up

Step 4: referral to psychiatry *U* = Urgent; *S* = Soon; *R* = Routine.

- High suicide risk—*U*
- Severe self-neglect—*U*
- Depression complicated by psychotic symptoms—*U*
- Depression complicated by significant psychiatric co-morbidity or psychosocial factors—*R/S*
- Inadequate response to multiple treatments—*R*

Safety-netting Follow up if patients do not attend appointments. Give patient ± family/carers clear advice on what to do if the patient's mood deteriorates and how to access urgent support, both in and out of hours.

Further information

NICE (2009) Depression in adults with a chronic physical health problem. 🖱 www.nice.org.uk/guidance/cg91

NICE (2009, updated 2018) Depression in adults: recognition and management. 🖱 www.nice.org.uk/guidance/cg90

Royal College of Psychiatrists (2015) Providing evidence-based psychological therapies to people with long-term conditions and/or medically unexplained symptoms. 🖱 https://www.rcpsych.ac.uk/pdf/PS02_2015.pdf

Patient information and support

Depression Alliance ☎ 0845 123 2320 🖱 www.depressionalliance.org
Samaritans ☎ 08457 909 090 🖱 www.samaritans.org

Drugs for treating depression

When should antidepressants be started? Consider for:

- Patients with moderate/severe depression ± psychological therapy
- Dysthymia (subthreshold depressive symptoms lasting >2y)
- Mild depression if other treatment strategies have failed—➔ p. 980

What should I tell the patient? Giving patients information ↑ compliance. When starting antidepressant drugs explain:

- The reasons for prescribing
- Timescale of action—immediate effect but unlikely to notice significant improvement for >2wk, maximum effect at 4–6wk
- Likely side effects including possible ↑ in anxiety in the first 2wk, and
- Likely duration of treatment: ≥6mo from recovery

⚠ Use of antidepressants has been associated with suicidal thoughts/behaviour. Those with a past history of self-harm and suicidal behaviour and children/young people are most at risk. Monitor for suicide risk especially at the start of treatment or after dose changes.

Which drugs are available? The major groups are:

Selective serotonin re-uptake inhibitors (SSRIs) e.g. fluoxetine 20–60mg od; sertraline 50–200mg od. Usually first choice as less likely to be discontinued due to side effects and safer in overdose. Warn of possible short-term ↑ in anxiety/agitation when starting (stop if significant). GI side effects including dyspepsia are common. Consider co-prescribing a PPI for stomach protection if >60y or other risk factors for GI bleeding.

Only fluoxetine has been shown to be of benefit for the treatment of depression in children—➔ p. 899

Elderly people, particularly those taking SSRIs, are prone to hyponatraemia when taking antidepressants.

Serotonin and noradrenaline reuptake inhibitors (SNRIs) e.g. venlafaxine 75–375mg/d, duloxetine 60–120mg/d. Avoid if uncontrolled hypertension. Venlafaxine is contraindicated if high risk of arrhythmia.

Tricyclic antidepressants (TCAs) e.g. lofepramine 70mg od/bd/tds, imipramine 10–300mg/d. Titrate dose until the patient feels the drug is helping *or* until side effects intrude. *Common side effects*: drowsiness, dry mouth, blurred vision, constipation, urinary retention, sweating. Use with caution if history of CVD (risk of arrhythmia), prostatic hypertrophy (↑ risk retention), and ↑ intraocular pressure (↑ risk acute glaucoma).

Vortioxetine Serotonin re-uptake inhibitor + antagonist of 5-HT$_3$ and agonist of 5-HT$_{1A}$ receptors. Anxiolytic/antidepressant. Dose: 5–20mg od. Option for treating major depression in adults who have responded inadequately to ≥2 antidepressants within the current episode.

Trazodone 100–600mg daily. Serotonin antagonist and reuptake inhibitor (SARI). Useful if sedation is needed. Avoid if history of arrhythmia/heart block, early after MI, or in the manic phase of bipolar disorder.

Mirtazapine 15–45mg nocte. Tetracyclic antidepressant. Presynaptic $\alpha 2$-adrenoreceptor antagonist. ↑ central noradrenergic and serotonergic neurotransmission. Causes sedation during initial treatment and may also cause weight ↑.

Reboxetine 4–6mg bd. Selective inhibitor of noradrenaline re-uptake. Not recommended for elderly patients.

Monoamine oxidase inhibitors (MAOIs) e.g. phenelzine 15mg tds. Should only be initiated in a specialist setting. Do not start until:

- >1–2wk after a tricyclic is stopped (3wk if clomipramine/imipramine)
- >1wk after an SSRI is stopped (2wk if sertraline; 5wk if fluoxetine)

Patients must be careful with diet, eating only fresh foods, and avoiding game, alcohol, and foods containing tyramine, e.g. mature cheese, pickled herring, broad bean pods, and meat, yeast, or soya bean extracts. Failure to do so results in rapid ↑ in BP (often preceded by a headache).

❗ Do not start other antidepressants until 2wk after treatment with MAOIs has been stopped (3wk for clomipramine/imipramine).

⚠ **Serotonin syndrome (serotonin toxicity)** Rare adverse drug reaction. Caused by excess serotonergic activity. Triggered by starting, ↑ dose, or overdose of a serotonergic drug; adding a new serotonergic drug (e.g. SSRI + TCA), or replacement of one serotonergic drug with another without a long enough washout period. Presents with:

- ↑ neuromuscular activity, e.g. tremor, ↑ reflexes, muscle twitching
- Autonomic dysfunction, e.g. tachycardia, BP changes, hyperthermia, sweating, shivering, diarrhoea, and/or
- Altered mental state, e.g. agitation, confusion, mania

Management Seek specialist advice. Usually involves stopping the involved medication and supportive care.

Follow-up Review patients every 1–2wk until stable assessing response, compliance, side effects, and suicidal risk. Continue for 4–6wk before judging a treatment has failed—and a further 2–4wk if partial response. Continue treatment for ≥6mo in total; ≥12mo in the elderly and those with GAD. If history of recurrent depression should continue for ≥2y.[N]

Discontinuation reactions Occur once a drug has been used ≥8wk. Usually apparent <5d after stopping the drug. Worse for drugs with a shorter half-life (e.g. paroxetine, venlafaxine). ↓ risk by tapering dose over ≥4wk (up to 6mo for patients on long-term maintenance therapy). Warn about possible reactions:

- SSRIs/SNRIs—GI disturbances, nausea, headache, paraesthesia, dizziness, anxiety, tinnitus, sleep disturbances, flu-like symptoms, sweating
- Other antidepressants (especially MAOIs)—nausea, vomiting, anorexia, headache, flu-like symptoms, insomnia, paraesthesiae, anxiety/panic and restlessness

St John's wort (*Hypericum perforatum*) Herbal remedy for treating mild depression. Do not prescribe/recommend as amount of active ingredient varies between formulations and has important interactions with other prescribed drugs, including antidepressants.

Psychosis

Characterized by loss of the link between reason and the outside world. Lifetime risk 3/100; 80% are aged <30y at diagnosis (5% <15y). Risk is ↑ by prolonged cannabis use and living in an inner city (3× ↑ risk).

Psychosis is not a diagnosis but a class of illnesses with 3 key features:

- Hallucinations—➔ p. 966
- Delusions—➔ p. 967
- Thought disorder -➔ p. 966

If ≥1 of these features is present diagnosis is very limited:

- **Affective psychosis** Psychotic depression, mania, and hypomania
- **Delusional psychosis** Schizophrenia/paranoid psychosis, *or*
- **Organic psychosis** Dementia; delirium

Schizophrenia ➔ p. 986 **Mania and hypomania** ➔ p. 987

Early intervention in psychosis Important to delay/prevent onset of disabling illness and ↓ complications (50% ↓ suicide rate; ↑ employment from 22% to 50%; ↓ social exclusion). Early intervention also ↓ healthcare costs by a third as a result of ↓ admissions.

Early symptoms Psychosis is usually heralded by gradual ↓ in intellectual/social functioning. ❗ Always take family concerns seriously.

- Poor sleep
- Panic
- Mood changes
- Social withdrawal/isolation
- Loss of job
- Broken relationships
- Undue suspicions/mistrust
- Perceptual changes

Seek evidence of psychotic thinking. Useful questions include:

- Do you think that you are different from other people?
- Do you think something strange is happening that you can't explain?
- Do you see, hear, feel, or experience things that others don't?
- Do you think others are watching you, talking about you, or having a go at you for no good reason?
- Do you think that you are special or important in some way?

Management If early psychosis is suspected, exclude other physical causes of symptoms (drug/alcohol abuse or withdrawal; metabolic abnormalities, e.g. thyrotoxicosis; hypoglycaemia; neurological disease, e.g. tumour, epilepsy; infection), and refer promptly to the early psychosis clinic for specialist assessment, diagnosis, and ongoing management.

❗ Alcohol/drug misuse is a common co-morbidity.

Physical health problems and psychosis People with psychotic illness die on average 16–25y early. Premature deaths result from:

- Suicide (33%)
- Physical disease (66%)—diabetes (2× ↑ prevalence), respiratory (10× ↑ mortality), cardiovascular (7× ↑ mortality), and infectious disease

Reasons

- Lifestyle—smoking (66% smoke); obesity (2–3×); poor diet, ↓ exercise
- Medication—some antipsychotic medications cause weight ↑ and adversely affect lipid profile; sedation may ↓ exercise
- Genetic—people with a diagnosis of psychosis are more likely to suffer from DM and/or CVD even after exclusion of other risk factors
- Social exclusion—↓ attendance at chronic disease management clinics, ↓ participation in screening programmes

Annual reviews in primary care All patients with confirmed non-organic psychotic illness require long-term follow-up in primary care.

- Regular reviews Case registers help ensure regular reviews take place. Check a written care plan has been drawn up by the psychiatric service involved. Follow up if a patient does not attend for review
- Check mental health Assess symptoms; compliance with medication; efficacy of treatment; medication side effects; risks of suicide
- Check physical health Annual screening and review of smoking, exercise, BMI (and/or waist circumference), BP, FBG or HbA1c, FBC, renal and liver function, lipids, prolactin (if taking antipsychotic medication), TFTs (if taking lithium)
- Education Involve family if possible—information about psychotic illness and treatment (reinforce compliance), early signs of relapse, where to access help, benefits of ↓ smoking, exercise, healthy diet, weight control, sensible drinking, and avoidance of illicit drugs, e.g. cannabis/amphetamines which exacerbate symptoms and ↑ relapse
- Help with lifestyle change e.g. referral to smoking cessation clinic, dietician or weight management programme; exercise prescription
- Social support Assistance may be needed with: carer support, finances, housing, employment, structured daily activity, transport, social networks. Those who can help include: social services, community mental health team, housing officer, disability/peer support workers

Driving and psychotic illness Advise patients to inform the DVLA. Driving should cease during the acute illness (all drivers) and until stable with insight for ≥3y (lorry/bus drivers).

⚠ Emergency management of severe psychosis

- Severe crises that endanger the individual or others are difficult to manage in primary care. Treatment in hospital may be required
- If unwilling to accept voluntary admission, use compulsory admission under the Mental Health Act (➔ p. 1104)
- Sedation while awaiting admission may be required—try oral medication first, e.g. lorazepam 1–2mg or chlorpromazine 50–100mg PO; if refused and severe agitation, consider lorazepam 1–2mg IM or chlorpromazine 50mg IM (↓ dose for elderly patients; avoid if the patient is epileptic, or has been drinking alcohol or taking barbiturates)

Further information

IRIS Initiative (2014) GP guidance: Emerging psychosis and young people. 🖱 http://iris-initiative.org.uk/resources/primary-care-resources/

NICE (2014) Psychosis and schizophrenia in adults: prevention and management. 🖱 www.nice.org.uk/guidance/cg178

Patient information and support

MIND ☎ 0300 123 3393 🖱 www.mind.org

Royal College of Psychiatrists Health advice leaflets. 🖱 https://www.rcpsych.ac.uk/healthadvice/atozindex.aspx

Schizophrenia and mania

Management of first presentation If psychotic illness is suspected, ask yourself whether the patient is a risk to self/others.

- If a risk, request emergency specialist assessment and/or acute admission voluntarily or under the Mental Health Act (➔ p. 1104)
- If no immediate risk, refer through the local rapid-access early intervention in psychosis service if referral criteria are met (➔ p. 984)

Physical health problems of people with psychosis ➔ p. 984

Driving and psychosis ➔ p. 985

Schizophrenia Frightening and disabling condition in which the sufferer is unable to distinguish his internal from the outside world. Lifetime prevalence ≈1%. Peak age of onset—♂ (15–25y); ♀ (25–35y).

First-rank symptoms Reliable markers in ~70% of patients. ≥1 symptom is suggestive of schizophrenia:

- Auditory hallucinations in the form of a commentary
- Hearing own thoughts spoken aloud
- Hearing voices referring to the patient, made in the third person
- Somatic hallucinations
- Thought broadcast, withdrawal, insertion, or interruption
- Delusional perception
- Feelings or actions experienced as made or influenced by external agents (passivity feelings)

First presentation of schizophrenia Typically a young person <35y with +ve symptoms (delusions, hallucinations, and/or thought disorder). May lack insight. Initial approach may come from a relative/friend.

- Ask about symptoms—in particular thoughts and perceptions
- Assess the patient's behaviour and appearance. Look for evidence of ↓ self-care, loss of affect, poverty of thought, and social withdrawal
- Try to elicit any history of drug misuse
- Ask friends/relatives present to tell you about the patient's behaviour

Differential diagnosis Illicit drugs, temporal lobe epilepsy, hypoglycaemia, delirium, dementia, affective disorder, personality disorder.

Chronic schizophrenia Thought disorder and −ve symptoms (poverty of thought, apathy, inactivity, lack of volition, social withdrawal, and loss of affect). Aim to treat the disease, prevent relapse and ↑ quality of life.

Antipsychotic medication Initiate only under specialist supervision. Commonly used drugs include: aripiprazole, amisulpride, olanzapine, quetiapine, clozapine. Choice depends on predominant symptoms and potential side effects. Safety concerns include:

- Weight ↑;
- Insulin resistance/DM
- Dyslipidaemia and CVD
- ↓ BP
- Blood dyscrasias
- Hyperprolactinaemia
- Sexual dysfunction
- Impaired temperature control
- Prolonged QT syndrome (check ECG before initiating)
- Extrapyramidal syndrome—parkinsonian symptoms; dystonia; restlessness; and tardive dyskinesia (➔ p. 522). Take specialist advice
- Neuroleptic malignant syndrome—hyperthermia, fluctuating consciousness, muscle rigidity, autonomic dysfunction (pallor, tachycardia, labile BP, sweating, urinary incontinence). Admit. Treatment is supportive—symptoms last 5–7d after drug is stopped

Mania and hypomania

Mania Characterized by a persistently high or euphoric mood (≥4d) out of keeping with circumstances. Other signs include:

- ↑ pressure of speech
- ↑ energy and activity
- ↑ appetite
- ↑ sexual desire
- ↑ pain threshold
- ↓ desire/need for sleep
- ↓ insight
- Grandiose delusions
- Hallucinations
- Labile mood
- Over-assertiveness
- Spending sprees
- Disinhibition
- Self-important ideas
- Poor concentration

Hypomania A less severe form of mania.

Differential diagnosis Hypoglycaemia; alcohol or drug abuse; prescribed drug side effects (e.g. steroids); temporal lobe epilepsy; frontal lobe dysfunction (e.g. due to tumour or stroke); thyrotoxicosis.

Lithium treatment Drug of choice—initiate only under specialist supervision. Check levels weekly until the dose is constant for 4wk, then monthly for 6mo; thereafter (as long as the dose is constant), check levels every 3mo. Check plasma creatinine, eGFR, and TFTs every 6mo. Keep fluid intake at roughly the same level each day. Avoid changing proprietary brands as bioavailability varies. *Toxicity*: blurred vision, diarrhoea/vomiting, ↓ K^+, drowsiness, ataxia, coarse tremor, dysarthria, hyperextension, fits, psychosis, coma, shock.

Alternative drugs Sodium valproate (teratogenic; avoid in women of child-bearing age—if essential, warn patients and ensure adequate contraception); carbamazepine.

Bipolar disorder or manic depression Consists of episodes when the patient has mania (bipolar I) or hypomania (bipolar II) against a background of depression. Lifetime prevalence ≈1%. ♂:♀ ≈1:1. *Peak incidence*: late teens and early 20s (90% are <30y). Management is as for mania.

Regular primary care review ➲ p. 985. ❗ All patients taking antipsychotic agents should have annual review of BMI, BP, FBG/HbA1c, lipids, FBC, U&E and eGFR, LFTs, and prolactin.

Referral to the specialist mental health team *U* = Urgent; *S* = Soon; *R* = Routine.

- ↑ in risk to self or others—*U*
- Poor response to treatment/persistent symptoms—*U*/*S*/*R*
- Significant side effects of medication—*U*/*S*/*R*
- Problems with adherence to treatment regimen—*S*/*R*
- Suspected co-morbid substance misuse—*R*

Further information

NICE (2014) Psychosis and schizophrenia in adults: prevention and management. 🖱 www.nice.org.uk/guidance/cg178

NICE (2014, updated 2018) Bipolar disorder: assessment and management. 🖱 www.nice.org.uk/guidance/cg185

Patient information and support

Bipolar UK ☎ 0333 3233 3880 🖱 www.bipolaruk.org
MIND ☎ 0300 123 3393 🖱 www.mind.org.uk
Rethink ☎ 0300 5000 927 🖱 www.rethink.org

Acute delirium

Common condition seen in general practice—particularly among elderly patients. May occur *de novo* or be superimposed upon chronic confusion of dementia (➔ p. 990) causing sudden worsening of cognition.

Presentation

- Global cognitive deficit with onset over hours/days
- Fluctuating conscious level—typically worse at night/late afternoon
- Impaired memory—on recovery amnesia of the events is usual
- Disorientation in time and place
- Odd behaviour—may be underactive, drowsy, and/or withdrawn *or* hyperactive and agitated
- Disordered thinking—often slow and muddled ± delusions (e.g. accuse relatives of taking things)
- Disturbed perceptions—hallucinations (particularly visual) are common
- Mood swings

Examination Can be difficult. If possible, do a thorough general physical examination to exclude treatable causes.

Assessment of capacity to make decisions ➔ p. 96

Possible causes

- Infection Particularly UTI, pneumonia; rarely encephalitis/meningitis
- Drugs Opioids, sedatives, levodopa, anticonvulsants, recreational drugs
- Metabolic Hypoglycaemia, hyponatraemia, uraemia, liver failure, hypercalcaemia, other electrolyte imbalance (rarer)
- Alcohol or drug withdrawal
- Hypoxia e.g. pneumonia, exacerbation of COPD, cardiac failure
- Cardiovascular MI, stroke, TIA
- Intracranial lesion Space-occupying lesion, ↑ ICP, head injury (especially subdural haematoma)
- Thyroid disease
- Carcinomatosis
- Epilepsy Temporal lobe epilepsy, postictal state
- Nutritional deficiency Vitamin B_{12}, thiamine, or nicotinic acid deficiency

Differential diagnosis

- Deafness May appear confused
- Dementia Longer history and lack of fluctuations in conscious level—in practice may be difficult to distinguish especially if you come across a patient who is alone and can give no history
- Primary mental illness e.g. schizophrenia (➔ p. 986); mania (➔ p. 987); anxiety state (➔ p. 970)

Management Is aimed at treating all remediable causes.

Admit to hospital or refer to intermediate care if

- The patient lives alone
- The patient will be left unsupervised for any duration of time
- If carers (or residential home) are unprepared/unable to continue looking after the patient, *and/or*
- If history/examination have indicated a cause requiring hospitalization

Possible investigations to consider in the community

- Cognitive function test, e.g. 6CIT (Table 26.1)
- Urine—dipstick for glucose, ketones, blood, protein, nitrates, and leucocyte esterase; send for M,C&S
- Check finger-prick capillary glucose to exclude hypoglycaemia
- Blood—FBC, ESR, U&E, eGFR, LFTs, Ca^{2+}, TFTs
- ECG
- CXR

Management at home

- Acute confusion is frightening for carers—reassure and support them
- Treat the cause, e.g. antibiotics for UTI or chest infection
- Try to avoid sedation as this can make confusion worse. Where unavoidable, use lorazepam 0.5–1mg po/IM prn (maximum 4mg/24h)
- Involve district nursing services and/or intermediate care services, e.g. to provide incontinence aids, nursing care, additional support
- If cause does not become clear despite investigation, or the patient fails to improve with treatment, admit for further investigation and assessment

Further information

NICE (2010, updated 2019) Delirium: prevention, diagnosis and management. 🖱 www.nice.org.uk/guidance/cg103

Table 26.1 The 6 Cognitive Impairment Test (6CIT)

	Question	Response	Score
1.	*What year is it?*	Correct: 0; Incorrect: 4	
2.	*What month is it?*	Correct: 0; Incorrect: 3	
Remember the following address: e.g. John Brown, 42, West Street, Bedford			
3.	*What time is it (to the nearest hour)?*	Correct: 0; Incorrect: 3	
4.	*Count backwards from 20 to1*	Correct: 0; 1 error: 2; >1 error: 4	
5.	*Months of the year backwards*	Correct: 0; 1 error: 2; >1 error: 4	
6.	*Repeat the memory phrase*	Correct: 0; 1 error: 2; 2 errors: 4; 3 errors: 6; 4 errors: 8; All incorrect: 10	
	Total		

Instructions on scoring Ring the appropriate score results for each question; add up the scores to produce a result out of 28.

Score

0–7	Not significant
8–9	Probably significant—refer—possible dementia
10–28	Significant—refer—likely dementia

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Dementia

Generalized impairment of intellect, memory, and personality, with no impairment of consciousness. *Prevalence* ↑ with age (rare <60y; 5% >65y; 20% >80y). *Common causes*: Alzheimer's disease (60%); vascular (multi-infarct) dementia; dementia with Lewy bodies.

Presentation Patients may be aware of 'being a bit forgetful' but usually relatives complain about their behaviour. Early symptoms are loss of short-term memory and inability to perform normally simple tasks. Alternatively, patients present later with failure to cope at home or self-neglect, occasionally leading to crisis. To diagnose dementia there must be a clear history of progressive impairment of memory and cognition ± personality change. Always assess level of support in the home, housing, and ability to cope (both patient and carers). Review medications to identify any that might impair cognition.

Examination Check general appearance—look for self-neglect, malnutrition, abuse; screen for cognitive deficit, e.g. with 6CIT (➔ p. 989).

Investigation Aimed at detecting treatable causes: check FBC, U&E, eGFR, LFTs, Ca^{2+}, TFTs, glucose, vitamin B_{12}, folate. Consider: MSU, CXR, ECG.

Differential diagnosis

- Acute delirium—➔ p. 988
- Depression—➔ p. 978
- Communication difficulties—deafness, dysphasia, or language difficulties

Prevention[N]

- 2° prevention: review and treat vascular risk factors
- Offer referral to genetic counselling to those thought to have a genetic cause for their dementia, and refer unaffected relatives

General management

- **Refer** All patients should be referred to the mental health services for the elderly or a memory assessment clinic for formal diagnosis, exclusion of treatable causes, ongoing specialist support and assessment, care planning, and community support
- **Apply principles of rehabilitation** ➔ p. 196; ➔ p. 554
- **Support carers** (➔ p. 200). Advise regarding benefits (➔ p. 108), self-help groups, respite care. Warn that dementia is progressive and prepare carers for a time when the patient does not recognize them
- **Discuss while sufferer still has capacity** Along with carers, the use of advance statements, lasting power of attorney (➔ p. 96), advanced decisions to refuse treatment, and preferred place of care plans
- **Treat concurrent problems** (e.g. UTI, chest infection, anaemia, pain) May make dementia worse. Consider possible side effects of medication. ❗ 40% have concurrent depression
- **Management of memory loss** Notebook to record 'tasks must do'; electronic prompts on mobile phone; medication dispensers

Management of behavioural (non-cognitive) symptoms

- Maintain a constant environment if possible
- Safety—arrange for door catches to prevent wandering and take up loose carpets to prevent falls; consider fire and electrical safety

- Avoid sedatives wherever possible as may worsen confusion—if needed use very low dose and review regularly

⚠ **Antipsychotic drugs for patients with dementia**[N] Do not use antipsychotics for mild–moderate non-cognitive symptoms because of the risk of severe adverse reaction, CVA (2× ↑ risk), and death. Consider in severe agitation only if:

- Risks and benefits have been assessed and discussed
- Patient is carefully monitored for changes in cognition
- Co-morbid conditions such as depression have been considered
- Dose is started low and titrated upwards
- Treatment is time limited and regularly reviewed (maximum 3mo)

Driving and dementia Group 1 licence (car/motorcycle)—patients should inform the DVLA. In early dementia, if sufficient skills are retained and progression is slow, a licence may be issued subject to annual review. Group 2 licence (bus/lorry)—licence revoked.

Assessment of capacity to make decisions ➔ p. 96

Alzheimer's dementia Most common form of dementia. *Cause*: unknown—defective genes found on chromosomes 14, 19, and 21. *Risk factors*: FH, Down's syndrome (onset at ~30y), late-onset depression, hypothyroidism, history of head injury. Presents with steady ↓ in memory and cognition. *Onset*: any age—normally >40y. ♀:♂ ≈0.7.

Drug management of cognitive symptoms Specialist initiated—refer. Preferred drug varies according to severity of cognitive deficit.

- Mild/moderate Anticholinesterase inhibitors (e.g. donepezil, galantamine, rivastigmine) ↓ rate of decline
- Moderate/severe Memantine ↓ clinical/cognitive decline

Vascular (multi-infarct) dementia Multiple lacunar infarcts or larger strokes cause generalized intellectual impairment. Tends to occur in a stepwise progression with each subsequent infarct. The final picture is one of dementia, pseudobulbar palsy, and shuffling gait with small steps. Treatment is as for secondary prevention of TIA/stroke—➔ p. 536.

Lewy body dementia Fluctuating but persistent cognitive impairment, parkinsonism, and hallucinations. No specific treatment. Avoid antipsychotics as they can be fatal. Use benzodiazepines if tranquilization is necessary.

Pick dementia Dementia characterized by personality change associated with frontal lobe signs such as gross tactlessness. Lack of restraint may lead to stealing, practical jokes, and unusual sexual adventures. Treatment is supportive.

Further information

NICE (2018) Dementia: assessment, management and support for people living with dementia and their carers. 🖱 www.nice.org.uk/guidance/ng97

Patient information and support

Alzheimer's Society ☎ 0300 222 1122 🖱 www.alzheimers.org.uk
Carers UK ☎ 0808 808 7777 🖱 www.carersuk.org

Eating disorders

Identification of and screening for eating disorders Target groups for screening include:

- Young women with low BMI compared with age norms
- Patients consulting with weight concerns who are not overweight
- Women with menstrual disturbances or amenorrhoea
- Patients with GI symptoms
- Patients with symptoms/signs of starvation—sensitivity to cold, delayed gastric emptying, constipation, ↓ BP, bradycardia, hypothermia
- Patients with physical signs of repeated vomiting—pitted teeth ± dental caries, general weakness, cardiac arrhythmias, renal damage, ↑ risk of UTI, epileptic fits, ↓ K^+
- Children with poor growth
- Young people with type 1 DM and poor treatment adherence

Screen target populations with the SCOFF questionnaire (Box 26.2).

⚠ Patients who are pregnant or have DM are at particular risk of complications if they have eating disorders. Refer early for specialist support and ensure everyone involved in care is aware of the eating disorder.

Anorexia nervosa Prevalence 0.02–0.04%. ♀ >> ♂. Usually begins in adolescence. Peak prevalence at 16–17y. *Features*:

- Refusal to keep body weight >85% of that expected (BMI <17.5kg/m^2)
- Intense fear of gaining weight, though underweight
- Disturbed experience of body weight or shape or undue influence of shape on self-image
- Amenorrhoea in women for ≥3mo and ↓ sexual interest

Patients tend to have a set daily calorific intake, e.g. 600–1000 calories and may employ strategies, e.g. bingeing and vomiting, purging, or excessive exercise to try to lose weight. Depression and social withdrawal are common as are symptoms 2° to starvation (see earlier 'Identification and screening for eating disorders' topic).

Management^N

- Give ongoing support and information
- Check electrolytes
- Refer to a specialist eating disorders clinic (if available) or the mental health team. Treatment involves family therapy for adolescents, psychotherapy, and possible admission for refeeding

Follow-up Patients with enduring anorexia nervosa not under 2° care follow-up should be offered an annual physical and mental health check.

⚠ Many patients with anorexia have compromised cardiac function. Avoid prescribing drugs adversely affecting cardiac function (e.g. antipsychotics, TCAs, macrolide antibiotics, some antihistamines). If prescribing is essential, then do a baseline ECG and follow-up ECG monitoring

Bulimia nervosa Prevalence 1–2%. Mainly ♀ aged 16–40y. *Features*:

- Self-image unduly influenced by body shape
- Recurrent episodes of binge eating, far beyond normally accepted amounts of food

- Inappropriate compensatory behaviour to prevent weight ↑, e.g. vomiting; use of laxatives, diuretics, and/or appetite suppressants. Bulimics can be subdivided into those that purge and those that just use fasting and exercise to control their weight
- Normal menses and normal weight. If low BMI, classified as anorexia

Management

- Give ongoing support and information
- Check electrolytes
- First-line treatments—evidence-based self-help, e.g. 'Self-help for bulimia and binge eating' available from 🖱 www.get.gg/bulimia.htm ± antidepressant medication (fluoxetine 60mg od)
- If unsuccessful, refer to a specialist eating disorders clinic (if available) or the mental health team. CBT may help

Advice for patients purging

- **Vomiting** Advise patients to avoid brushing their teeth after vomiting, rinse with a non-acid mouthwash after vomiting, and ↓ acid oral environment (e.g. by limiting acid food and drink)
- **Laxatives** If laxative abuse, advise patients that laxatives do not significantly ↓ calorie absorption and to gradually ↓ laxative intake

Binge eating disorder Pattern of consumption of large amounts of food, even when not hungry. Common. Usually associated with obsessive feelings about food and body image, feelings of guilt/disgust about the amounts consumed, and/or a feeling of lack of control. *Management*:

- Give ongoing support and information
- Provide an evidence-based self-help programme as a first step and/or antidepressant medication (SSRI is the drug group of choice)
- If unsuccessful refer for specialist help. CBT might be helpful
- In all cases, provide concurrent advice and support to tackle any co-morbid obesity (➔ p. 152)

Body dysmorphic disorder ➔ p. 975

Box 26.2 The SCOFF questionnaire

- Do you make yourself Sick because you feel uncomfortably full?
- Do you worry you have lost Control over how much you eat?
- Have you recently lost One stone in a 3mo period?
- Do you believe yourself to be Fat when others say you are too thin?
- Would you say that Food dominates your life?

Score 1 point for each +ve answer; a score of ≥2 suggests anorexia/bulimia is likely.

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Further information

NICE (2017) Eating disorders: recognition and treatment. 🖱 https://www.nice.org.uk/guidance/ng69

Patient support and information

Beating Eating Disorders (BEAT) ☎ 0800 801 0677 (adults); ☎ 0800 801 0711 (youths) 🖱 www.b-eat.co.uk

Other psychological conditions

Seasonal affective disorder (SAD) 'Winter blues'—recurrent disorder involving 'seasonal' episodes of depression, usually in the winter months. Affects ≈2% adults. ♀:♂ ≈2:1. Peak incidence third decade.

Symptoms Depression + ↑ sleep, ↑ food intake (with carbohydrate craving), and weight ↑. 30% experience elatory mood swings in summer.

Management SSRIs (particularly sertraline); phototherapy (30–90min/d in the early morning—effects should be seen within 3wk). Light boxes can be borrowed from psychiatry departments, hired, or bought (contact SAD Association for more information 🖱 www.sada.org.uk).

Emotionally unstable (borderline) personality disorder Affects ~1% of the population. ♀ > ♂. *Features*:

- Pervasive maladaptive patterns of behaviour, thinking, and control of emotions
- Significant instability of personal relationships, self-image, and mood
- Impulsive behaviour
- Chronic feelings of emptiness
- Tendency towards suicidal thoughts/self-harm
- Possible transient psychotic symptoms

Recognition Consider if repeated self-harm, persistent risk-taking, or marked emotional instability. If suspected, refer for specialist assessment.

Crisis management[N]

- Assess current risk to self/others—if a risk, request emergency specialist assessment and/or acute admission voluntarily or under the Mental Health Act (➔ p. 1104)
- Ask about past episodes and management strategies that worked
- Help to identify manageable changes that might enable the patient to deal with current problems
- Offer follow-up and contact the patient if the appointment is not kept
- Refer for specialist care if risk/distress continues to ↑

Specialist treatment of personality disorder

- Intensive structured psychological therapy, e.g. STEPPS programme
- Drugs—SSRIs are used for impulsive behaviour; low-dose antipsychotics for paranoid ideas; mood stabilizers for emotional instability. ❗ Be careful taking over prescribing from specialist care—no drugs are licensed for personality disorder and evidence of efficacy is poor

⚠ Risk of overdose is ↑ among patients with personality disorder.

Factitious disorder (Munchausen syndrome) Intentional feigning of physical/psychological symptoms to assume the sick role (± hospital admission). Can be difficult to detect. Differs from malingering as there is no external reward (e.g. financial). *Common presentations*:

- Physical Dermatitis artefacta, PUO, bruising disorders, brittle DM, diarrhoea of unknown cause; neurological symptoms, e.g. pseudoparalysis or pseudofits (neurologica diabolica); abdominal pain (laparotomophilia migrans); chest pain (cardiopathia fantastica)
- Psychological Feigned psychosis, fictitious bereavement, fictitious overdose

Management Exclude any other basis for presenting pathology. Explain findings to the patient exploring possible causes. Assess psychological and social difficulties. Consider referral to specialist mental health services.

Munchausen syndrome by proxy Caregiver—typically a mother with child—seeks repeated medical investigations and needless treatment for the person he/she is caring for. The child or person being cared for may actually be harmed by the carer to achieve these aims. Commonly reported symptoms include: neurological symptoms, bleeding, rashes. Often difficult to detect and even harder to prove. A form of abuse that must be taken seriously and handled with care (➔ p. 902). Involve all relevant agencies early (e.g. social services, paediatrics).

Malingering Intentional production or feigning of physical or psychological symptoms to assume the sick role for a known external purpose. Malingering is not considered a mental illness or psychopathology, although it can occur in the context of other mental illnesses. Forms:

- Pure malingering The individual falsifies all symptoms
- Partial malingering The individual has symptoms but exaggerates the impact they have upon daily functioning
- Simulation The individual acts out symptoms of a specific disability
- False attribution The individual has valid symptoms but is dishonest as to the source of the problems, e.g. attributing neck pain to a RTA to obtain compensation

Differential diagnosis True medical or psychiatric illness yet to be diagnosed; factitious disorder; somatization disorder.

Common motivating factors

- Avoidance of work and/or other responsibility
- Litigation to obtain money
- Obtaining narcotics
- Avoidance of/release from jail
- Need for attention

Management Difficult. As doctors we tend to believe our patients.

- Exclude causes for symptoms through careful history/examination
- Avoid prescribing drugs and unnecessary referrals as these might perpetuate symptoms
- Avoid certifying the patient as unfit to work or perform activities—if the patient is unhappy about this, suggest a second opinion
- Tactfully explain your findings and conclusions to the patient and explore the reasons for the behaviour
- Provide support to find more appropriate ways to solve problems

Further information

NICE (2009) Borderline personality disorder. 🖱 www.nice.org.uk/guidance/cg78

Patient/relative information and support

Borderline personality disorder (BPD) Central 🖱 www.bpdcentral.com
Emergence 🖱 www.emergenceplus.org.uk
Royal College of Psychiatrists (2016) Self-harm. 🖱 www.rcpsych.ac.uk/healthadvice/problemsdisorders/self-harm.aspx

Chapter 27

Cancer care

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Surgery for cancer *1004*
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Diagnosis of cancer

Genetic predisposition to cancer Identifying people with an inherited tendency to cancer development can enable screening for early detection and/or preventive measures. Consider if family history of cancer in multiple family members, cancer at unusually young age in another family member, or certain congenital genetic syndromes. *Examples*:

- **Congenital syndromes** Tuberous sclerosis (renal cancer); Down's syndrome (leukaemia); neurofibromatosis (sarcoma, brain tumour, leukaemia, breast and colorectal cancer)
- **Breast cancer** *BRCA1/2, TP53, PALB2*, and *PTEN* genes (➔ p. 673)
- **Bowel cancer** Lynch syndrome (*MLH1, MSH2/6*, and *PMS2* genes); familial adenomatous polyposis (*APC* gene); Peutz–Jeghers syndrome (*STK11* gene); juvenile polyposis (*BMPR1A/SMAD4* genes)—➔ p. 368

Referral guidelines for suspected cancer To promote earlier detection and ↑ survival rates, the threshold for referral for investigation of suspected cancer is set at a positive predictive value of 3% (Table 27.1). In all cases, patients should be referred to be seen by an appropriate specialist team in <2wk. ❗ Referral guidance slightly differs in Scotland.

Before referral Explain to patients why they are being referred and discuss with them their likelihood of cancer and alternative diagnoses. Ensure patients are aware of support they can get while they await their appointment. Always check address and phone details are correct, and state in the referral letter whether an interpreter is needed.

Blood tests It is good practice to ask the patient to have baseline blood tests (e.g. FBC, eGFR, LFTs, FBG/HbA1c) done before their specialist appointment to avoid delaying specialist investigations.

Table 27.1 Summary of current NICE cancer referral guidelines[N]

Cancer type	Referral guidance
Brain and CNS ➔ p. 532	**Urgent direct access MRI** If progressive, sub-acute loss of CNS function **Same-day referral** If unexplained new cerebellar or other CNS symptoms
Lung ➔ p. 290 ❗ Normal CXR does not exclude lung cancer—refer if concerned	**Urgent CXR** If >40y and ≥1 (if smoker) or ≥2 (if non-smoker) of the following symptoms: • Chest pain • Weight loss • Anorexia • Cough • Fatigue • Dyspnoea
	Urgent CXR If >40y with: • Persistent/recurrent chest infection • Finger clubbing • Supraclavicular or persistent cervical LNs • Focal chest signs suggesting malignancy or pleural disease • Thrombocytosis **Urgent specialist referral** If >40y and unexplained haemoptysis
Larynx ➔ p. 914	**Urgent specialist referral** If >45y with: • Persistent unexplained hoarseness • Unexplained neck lump

(*Continued*)

Table 27.1 (*Contd.*)

Cancer type	Referral guidance
Thyroid ➔ p. 332	**Urgent specialist referral** If unexplained thyroid lump
Mouth ➔ p. 908	**Urgent specialist referral** If: • Lump on lip or oral cavity suggestive of oral cancer • Unexplained ulceration in the oral cavity lasting >21d • Persistent and unexplained neck lump • Red or white patch in oral cavity consistent with erythroplakia or erythroleucoplakia
Oesophageal and gastric ➔ p. 362	**Urgent specialist referral** If abdominal mass suggestive of gastric cancer **Urgent direct access OGD** If: • Dysphagia or • >55y and weight loss + abdominal pain/reflux/dyspepsia **Consider routine OGD** If: • Treatment-resistant dyspepsia • Upper abdominal pain + anaemia • Thrombocytosis + persistent nausea/vomiting, weight loss, reflux/dyspepsia, and/or upper abdominal pain • Nausea and vomiting + any of weight loss, reflux/dyspepsia, or upper abdominal pain
Gallbladder/ liver ➔ p. 400, p. 398, p. 399	**Urgent USS** If abdominal mass consistent with enlarged gallbladder or liver
Pancreatic ➔ p. 404	**Urgent specialist referral** If >40y with jaundice **Urgent direct access CT** If >60y with weight loss and: • Diarrhoea • Back pain • Abdominal pain • Persistent nausea/vomiting • Constipation • New-onset diabetes
Colorectal ➔ p. 370	**Urgent specialist referral** If: • >40y with unexplained weight ↓ + abdominal pain • >50y with unexplained rectal bleeding • >60y with iron deficiency anaemia or change in bowel habit • +ve faecal occult blood test • Rectal or abdominal mass consistent with colorectal cancer • Unexplained anal mass/ulceration • <50y with rectal bleeding + abdominal pain, change in bowel habit, weight ↓, or iron deficiency anaemia **Faecal occult blood testing** If no rectal bleeding but: • >50y with unexplained abdominal pain or weight ↓ (if both then immediate urgent referral) • <60y with change in bowel habit or iron deficiency anaemia (if both, then immediate urgent referral) • <60y with anaemia even in the absence of iron deficiency
Bladder and kidney ➔ p. 420	**Urgent specialist referral** If: • >45y and unexplained visible haematuria without UTI or haematuria that persists after treatment of UTI • >60y with unexplained non-visible haematuria + dysuria or leucocytosis

(*Continued*)

Table 27.1 (*Contd.*)

Cancer type	Referral guidance
Prostate ➔ p. 434	Consider non-urgent urology referral If >60y and recurrent or persistent UTI DRE + prostate-specific antigen (PSA) test If: • Lower urinary tract symptoms, e.g. nocturia, urinary frequency, hesitancy, urgency, or retention • Erectile dysfunction • Visible haematuria (if UTI excluded or persists after treated) Urgent specialist referral If: • Prostate feels malignant on DRE • PSA is above age-specific reference range
Testis ➔ p. 442	Urgent specialist referral If non-painful enlargement or change in shape or texture of the testis Consider USS If unexplained testicular symptoms which do not meet referral criteria
Penis ➔ p. 439	Urgent specialist referral If penile mass or ulceration and STI excluded or persistent penile lesion despite treatment for STI
Cervix, vulva, and vagina ➔ p. 702 & p. 706	Urgent specialist referral If clinical examination suggests malignancy
Ovary ➔ p. 699	Urgent specialist referral If clinical examination reveals ascites associated with a pelvic mass that is not obviously a fibroid Consider urgent CA 125 and/or urgent USS If any of the following symptoms are persistent/frequent (>12×/mo), especially if the women is >50y: • Persistent abdominal distention/bloating • Early satiety/loss of appetite • Pelvic/abdominal pain • ↑ urinary frequency/urgency • Unexplained weight loss • Fatigue • Change in bowel habit • New-onset irritable bowel syndrome in the past 12mo
Endometrium ➔ p. 697	Urgent specialist referral If postmenopausal bleeding. Consider direct access USS if >55y with any of: • New, unexplained vaginal discharge • Thrombocytosis • Visible haematuria • Anaemia and/or ↑ blood glucose
Breast ➔ p. 674	Urgent specialist referral If: • Skin changes suggestive of cancer, e.g. peau d'orange • >30y and unexplained breast or axillary lump • >50y with any of the following symptoms in one nipple: discharge, retraction, Paget's disease, or other skin changes
Skin: basal cell cancer (BCC) ➔ p. 604	Refer routinely Lesions with clinical features of BCC—typically raised ulcer with a rolled edge, blood vessels around edge, or pearly waxy nodule Consider urgent specialist referral If BCC is on a site or is of a size that delay in referral would have an unfavourable impact

(*Continued*)

Table 27.1 (*Contd.*)

Cancer type	Referral guidance	
Skin: squamous cell cancer (SCC) ➔ p. 604	Urgent specialist referral If skin lesion is suggestive of SCC, i.e. • Typically on sun-exposed site, e.g. head, neck, hand • Raised, ulcerated, keratinized, or crusting lesion	
Skin: melanoma ➔ p. 602	Urgent specialist referral If: • Clinical examination suggests nodular melanoma • Dermoscopy suggests malignant melanoma • A suspicious pigmented lesion score ≥3 from the following 7-point checklist	
	Major features (score 2 points) • Change in size • Irregular shape • Irregular colour	*Minor features* (score 1 point) • Largest diameter ≥7mm • Inflammation • Ooze • Change in sensation
Leukaemia ➔ p. 652	Offer FBC in <48h If: • Pallor • Persistent fatigue • Unexplained bruising or bleeding • Unexplained fever • Unexplained persistent infections • Generalized lymphadenopthy	*Adults only:* • Recurrent infections • Petechiae *Children only:* • Persistent or unexplained bone pain
	Same-day specialist review If ***child*** with unexplained petechiae or hepatosplenomegaly	
Lymphoma ➔ p. 656	In all cases, ask about fever, night sweats, shortness of breath, pruritus, or weight loss. Where appropriate, ask if symptoms are alcohol-related In unexplained lymphadenopathy or splenomegaly: • Adults Urgent specialist referral • Children and young people Admit or arrange specialist assessment in <48h	
Myeloma ➔ p. 650	Offer blood screen (FBC, serum calcium, plasma viscosity/ESR) If >60y and persistent bone pain—particularly back pain or unexplained fractures Offer protein electrophoresis and urine Bence Jones protein test in <48h If symptoms suggestive of myeloma and: • >60y with hypercalcaemia or leucopenia, or • Any age with raised plasma viscosity/ESR Urgent specialist referral If protein electrophoresis suggests myeloma	

Childhood cancers ➔ p. 884

Further information

NHS Scotland Scottish referral guidelines for suspected cancer. 🖱 www.cancerreferral.scot.nhs.uk

NICE (2015, updated 2017) Suspected cancer: recognition and referral. 🖱 www.nice.org.uk/guidance/ng12

Principles of cancer care

Treatment options If cancer is confirmed, comprehensive assessment is needed to make treatment decisions. Treatment can be:

- **Radical** Curative intent—surgery and/or drug/radiotherapy
- **Adjuvant** Given after surgery when micrometastatic disease is suspected; decision to proceed is based on likelihood of relapse
- **Neoadjuvant** Given prior to definitive treatment to make treatment easier and more likely to succeed
- **Palliative** When cure is not possible; symptom management is the priority—➔ p. 1011

Assessment of the tumour

- **Histological nature** Tissue of origin; cancer type (e.g. adenocarcinoma, squamous cell); degree of differentiation; and tumour grade. High-grade, poorly differentiated tumours tend to have a poorer outcome
- **Biological behaviour** Tumour markers produced by cancers may be a useful adjunct to histological classification and staging and can be used to influence and monitor efficacy of treatment (Table 27.2)
- **Genetic characteristics** Assessment of acquired gene changes in tumour cells may inform diagnosis, prognosis, and treatment options. For example: *JAK2* mutations inform diagnosis of myeloproliferative disorders and targeted treatments are being developed; the *BCR-ABL* fusion gene ('Philadelphia chromosome') informs diagnosis of CML; *AR*-v7 predicts prostate cancer poorly responsive to anti-androgen therapy
- **Anatomical extent** Determined through clinical, radiological, biochemical, and surgical assessment. Routine blood tests (e.g. LFTs, bone profile) may also indicate the presence of metastases

Cancer staging Allows the plan of treatment to be made.

The TMN classification A widely used classification of tumours. Exact criteria for staging depend on the primary organ site:

- **T** Primary tumour—graded T_1–T_4 with increasing size of primary
- **N** Regional lymph nodes—graded from N_0 to N_3
- **M** Presence (M_1) or absence (M_0) of metastases

Stage grouping

- **Stage 1** Tumour is clinically confined to the primary organ. The lesion tends to be operable and completely resectable
- **Stage 2** Clinical examination shows evidence of local spread into surrounding tissue and first draining LNs. The lesion is operable and resectable but there is a higher risk of further spread of disease
- **Stage 3** Clinical examination reveals extensive primary tumour with fixation to deeper structures and local invasion. The lesion may not be operable and requires a combination of treatment modalities
- **Stage 4** Evidence of distant metastases beyond the site of origin. The primary site may be surgically inoperable

Other factors to consider in determining treatment

- **Patient's performance status** Establishes the general fitness of the patient for treatment. Tools include: the Eastern Co-operative

Oncology Group (ECOG) Performance Status Scale; WHO Scale; Karnofsky Performance Status; and Lansky Scale (for children)

- Mortality, morbidity, and efficacy of the treatment
- Patient preferences

Table 27.2 Biochemical tumour markers and associated conditions

Tumour marker	Associations	
	Malignant	*Non-malignant*
CEA	GI tract cancers (particularly colorectal cancer)	Cirrhosis Pancreatitis Smoking
CA 19-9	Colorectal cancer Pancreatic cancer	Cholestasis
CA 125	Ovarian cancer Breast cancer Hepatocellular cancer	Cirrhosis Pregnancy Peritonitis Endometriosis
AFP	Hepatocellular cancer Germ cell cancers (not pure seminoma)	Liver disease—hepatitis/cirrhosis Pregnancy Open neural tube defects
β-hCG	Germ cell cancers Choriocarcinoma and hydatidiform mole	Pregnancy
PSA	Prostate cancer	BPH Prostatitis/UTI Prostate instrumentation Acute urinary retention Physical exercise Old age
LDH	Germ cell tumours Lymphoma Leukaemia Melanoma Neuroblastoma	Myocardial infarction or stroke Haemolytic anaemia Infectious mononucleosis Liver disease Muscle disease
β-2-microglobulin (β2M)	Myeloma (high levels predict poor prognosis) CLL Some lymphomas	Viral infections e.g. HIV, CMV MS

The GP's role Treatment for cancer is increasingly successful but also increasingly complex. It is largely a specialist activity, but the role of the GP is important at this time, even if mainly supportive.

- Keep in touch with the family and up to date with treatment—provide support (e.g. advice on benefits/local services), preventive care (e.g. flu vaccination), and general medical care
- Liaise with the secondary care teams involved, and provide continuity if care is passed from one specialist team to another
- If the patient does not survive, provide ongoing support to family

Surgery for cancer

Surgery has 3 main roles in cancer management:

Diagnosis and staging Advances in imaging and laparoscopic techniques have dramatically reduced the number of patients requiring open surgery to confirm a cancer diagnosis. However, surgical staging remains important in:

- Breast cancer—'sentinel' axillary node biopsy is needed to accurately predict the state of nodal disease
- Ovarian cancer—tumour deposits on the peritoneal surface are poorly visualized with conventional imaging. Direct visualization is required using laparotomy or laparoscopy
- Certain abdominal malignancies—laparoscopic assessment of extent and spread of tumour may be performed prior to major resection

Curative surgery

Non-metastatic disease Surgery with curative intent is dependent on complete resection of the tumour with a margin of normal tissue. Local control of tumours with a propensity to spread to lymph nodes may be improved with resection of the draining group of nodes, e.g. vulval tumours. However, even if the tumour was completely resected, surgery can still fail to cure either as a result of:

- Development of metastatic disease from micro-metastatic deposits unidentifiable at the time of surgery, or
- Development of local relapse as a result of inadequate margins

Metastatic disease Surgery may be curative in a limited number of tumours with metastases. Circumstances in which curative surgery may be offered include:

- Isolated brain metastases from breast cancer with a long disease-free interval
- Liver metastases from colorectal cancer
- Pulmonary metastases from osteosarcoma or soft tissue sarcoma

Palliative surgery Surgery can be effective in achieving good symptom control during palliation. Life expectancy, performance status, and tumour progression should be considered before embarking on surgery. Ideally such decisions should be multidisciplinary and involve surgeons specialized in oncology and experienced in palliative management (Table 27.3).

Table 27.3 Situations in which palliative surgery should be considered

Situation	Comments
Cancers causing obstructive symptoms e.g. bowel, ovary, ureter, bronchus ❗ Most malignancy-associated bowel obstructions are functional, not anatomical	Surgery to relieve the obstruction may be warranted even if the underlying disease is incurable with locally advanced disease or distant metastases **Bowel obstruction** This occurs most commonly in patients with colonic or ovarian cancer **Oesophageal or bronchial obstruction** Laser therapy of an intraluminal mass may restore the lumen **Obstructive hydronephrosis** Nephrostomy or ureteric catheters may relieve the obstruction **Placement of a stent** May help relieve the symptoms of dysphagia, dyspnoea, jaundice, and large bowel obstruction
Fistulae	Fistulae, often arising as a result of pelvic tumours or as a side effect of radiotherapy, can be associated with distressing malodours and excessive discharge
Jaundice	**Radiological and/or endoscopic stent placement** Can relieve obstructive jaundice secondary to extrinsic pressure from lymph nodes on the biliary system or intrinsic pressure from cholangiocarcinoma or pancreatic carcinoma. **Surgical relief** by choledochoenterostomy, avoids the problems associated with stents and may be indicated in a small minority with excellent performance status and slowly growing disease
Spinal cord compression and brain tumours	Urgent referral for neurological assessment for decompressive surgery or vertebroplasty is indicated for confirmed spinal disease or operable brain tumours
Gastrointestinal bleeding	A wide range of endoscopic techniques have been developed to stop bleeding from benign and malignant causes including sclerotherapy with adrenaline, laser coagulation, and radiological embolization These techniques may avoid the need for major surgery in patients who have a limited life expectancy
Bone metastases	Prophylactic fixation of a long bone may reduce either pain and/or the risk of pathological fracture in patients with: ● Lesions in weight-bearing bones ● Destruction of >50% of the cortex ● Pain on weight bearing ● Lytic lesions In all cases fixation should be followed by radiotherapy to control growth and promote healing
Pain	If the expected morbidity of the procedure is low, surgical debulking of large, slowly growing tumours can reduce pain Neurosurgical approaches such as cordotomy are only rarely considered
Ascites	Drainage of ascites can improve breathlessness, pain, and appetite. Frequent drainage is often required as ascites quickly accumulates. Indwelling catheters can be used in selected patients

Chemotherapy

Chemotherapy is the use of chemical agents in the cure or palliation of malignant disease.

Drug groups Include:

- Antibiotics, e.g. bleomycin
- Alkylating agents, e.g. busulfan
- Antimetabolites, e.g. methotrexate, fluorouracil
- Alkaloids, e.g. vincristine
- Platinum derivatives—DNA intercalating agents, e.g. cisplatin
- Enzymes, e.g. asparaginase
- Hormones, e.g. sex hormones, corticosteroids
- Biological agents, e.g. interferon, monoclonal antibodies
- Others, e.g. hydroxycarbamide, retinoids

These agents work in a variety of ways to inhibit tumour growth and/or cause tumour cell damage. Normal cells may be damaged at the same time as tumour cells, resulting in the high levels of toxicity experienced by patients.

Choice of agent Depends on known drug activity, whether patient is enrolled in a trial, cost, and patient factors. It is a specialist decision.

Types of tumour Response to chemotherapy depends on type and grade of tumour being treated. Broadly tumours can be divided into:

- Those likely to respond Leukaemia, lymphoma (Hodgkin's and intermediate/high-grade non-Hodgkin's), testicular tumours, small cell lung cancer, embryonal tumours, choriocarcinoma, ovarian cancer, sarcoma, breast cancer, prostate cancer
- Those that may respond Low-grade non-Hodgkin's lymphoma, GI cancer, brain/CNS tumours, melanoma, bladder and uterine cancer
- Those unlikely to respond Non-small cell lung, renal, pancreatic, head and neck, cervical, and liver cancer

Combination chemotherapy Often different chemotherapeutic agents are combined to ↑ their chances of effect. Agents acting in different ways may potentiate each other's actions. Using combinations also ↓ the risk of resistance (if one agent does not have any effect, another may). Choosing agents with different side effect profiles reduces cumulative toxic effects.

Intermittent chemotherapy Particularly useful for cytotoxic drugs. Intermittent treatment exploits the difference in recovery between normal and malignant tissues. Gaps between cycles of treatment allow normal tissue (particularly the immune system) to recover, but the malignant tissue does not recover to such an extent (Figure 27.1). The population of malignant cells diminishes relative to normal cells with each cycle.

Adjuvant chemotherapy Given to prevent relapse after primary treatment of a non-metastatic tumour for which relapse rate is known to be high. An example is adjuvant chemotherapy for breast cancer.

⚠ *Neutropenic sepsis* ➔ p. 626

Cancer immunotherapy Treatments that modify the immune response to alter tumour growth.

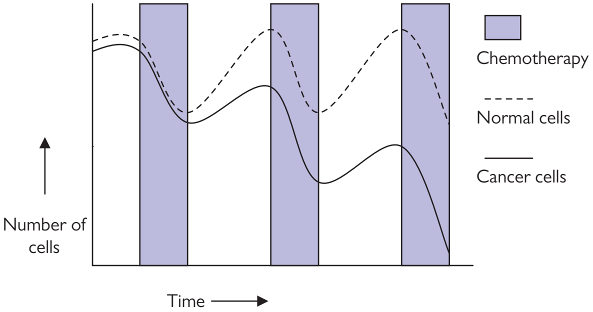

Figure 27.1 Action of cytotoxic chemotherapy on normal and cancer cell populations

[a] RTOG, Radiotherapy and Oncology Group.

Immune checkpoint proteins Utilized by some tumours to suppress the immune system thereby limiting the body's ability to repair. Certain drugs (e.g. ipilimumab—used in myeloma) block the activity of a checkpoint protein (in this case, CTLA4 expressed on T-lymphocytes) which enables T cells to destroy the cancer cells.

Adoptive cell transfer An experimental form of immune cell therapy. Cytokines on tumour infiltrating lymphocytes from the tumour itself are activated and reinfused into the patient's bloodstream to shrink or destroy the tumour.

Therapeutic antibodies Created by linking antibodies to a toxic substance which triggers cancer cell death (e.g. ado-trastuzumab emtansine in some forms of breast cancer). Other therapeutic antibiotics cause cancer cell apoptosis or immune-mediated cytotoxicity (e.g. rituximab for B-cell chronic lymphocytic leukaemia).

Cancer vaccines For some cancers, vaccines can be made using the patient's own tumour cells (e.g. sipuleucel-T in prostate cancer).

Immune system modulators Interleukins and interferons are cytokines which enhance the body's immune response to cancer. Examples of use include: renal cell cancer, melanoma, multiple myeloma, and some leukaemias. Side effects include fatigue, flu-like symptoms, diarrhoea, leucopenia, nausea, and anorexia.

Information for patients about side effects of chemotherapy

Cancer Research UK ☎ 0808 800 4040 🖱 www.cancerhelp.org.uk
Chemocare 🖱 www.chemocare.com
Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk

Radiotherapy

Mechanism of action Ionizing radiation damages cells. Radiotherapy aims to deliver a dose of irradiation to an area which allows normal tissues, but not the cancer, to recover from the damage.

Delivery of radiotherapy May be used alone or with chemotherapy. Once the maximum dose of radiotherapy has been received by any area, that area cannot usually be irradiated again.

- External beam External source of ionizing radiation (e.g. gamma rays) is aimed at a target point on the body. Patients may be immobilized (e.g. with boards/moulds) to ensure delivery to the correct place. Can be single dose (e.g. for palliative reasons) or fractionated into several doses spread over weeks. Fractionation ↑ effect
- Brachytherapy Delivery of radiation by placing a radioactive source within or close to the malignancy, e.g. caesium-137 in the uterus

Radiotherapy side effects Table 27.4 and Table 27.5

Table 27.4 Managing post-radiotherapy skin reactions

RTOG score	Description	Skin appearance	Treatment
0	Normal	Normal	Emollient bd to delay onset of reaction
1	Faint erythema	Skin slightly pink or red	Emollient tds or prn
2A	Tender or bright erythema (dry desquamation)	Skin red, dry, and scaly—some itch and tingling	Emollient (qds or prn) Hydrocortisone cream may be used sparingly on itchy areas. Review use after 7d—discontinue if the skin breaks down
2B	Patchy, moist desquamation, oedema	Skin inflamed with patches of epidermis broken down and moist	Consider involvement of district nursing team for dressing management (e.g. hydrogel dressing, surgipad, or foam dressing) Continue with emollient use
3	Confluent moist desquamation	Epidermis blisters and sloughs, underlying dermis is exposed and sore. Oozing of serous fluid	Consider involvement of district nursing team for dressing management (e.g. hydrogel dressing, surgipad, or foam dressing) Review frequently Swab and treat with oral antibiotics (e.g. flucloxacillin 500mg qds) if any signs of infection.
After radiotherapy	Reaction may continue for several weeks post treatment. Continue with use of emollients until skin returns to normal Continue involvement of district nursing team		

Radiotherapy treatment may be

- **Curative** e.g. childhood tumours, lymphoma, seminoma, head/neck tumours, bladder cancer, squamous/basal cell skin cancer
- **Adjuvant** Preoperatively to ↓ size/extent of otherwise inoperable tumours, or postoperatively to treat microscopic foci remaining after tumour removal (e.g. in treatment of breast cancer)
- **Palliative** For control of distressing symptoms. Only symptomatic sites are targeted, e.g. bone metastases; haemorrhage; obstruction of a viscus; neurological complications; fungating tumours

Table 27.5 Non-skin side effects of radiotherapy

Side effect	Description/action
Sore mouth/throat	Associated with head/neck radiotherapy. Advise to visit the dentist prior to treatment; avoid smoking, alcohol, and spicy foods; rest voice when radiotherapy reaction is established *Consider treatment with*: normal saline/bicarbonate mouthwashes; antiseptic mouth washes (e.g. chlorhexidine—though alcohol may sting); soluble aspirin (can be gargled) or paracetamol; benzydamine mouthwash; topical local anaesthetics (e.g. lidocaine® gel); topical steroids; coating agents (e.g. sucralfate). If insufficient fluid/food intake, consider nutritional support via nasogastric tube and/or referral for gastrostomy (if weight loss >10%)
Dysphagia	May result from thoracic radiotherapy. Avoid smoking, spirits, and spicy food. Consider treatment with: antacid; sucralfate; soluble paracetamol or aspirin; NSAID po/PR
Nausea and vomiting	Radiotherapy to the abdomen often causes nausea as a result of serotonin release. Consider prophylactic antiemetic therapy with a serotonin inhibitor, e.g. ondansetron
Diarrhoea	Frequently accompanies abdominal/pelvic radiotherapy *Management*: dietary modification (e.g. ↓ dietary fibre) may help. Supply with loperamide—4mg initial dose then 2mg every 2h until symptoms settle (4mg every 4h at night) *Proctitis* may accompany rectal/prostatic irradiation. Treat with rectal steroids
Pneumonitis	Acute pneumonitis can develop 1–3mo after treatment and is associated with a fever, dry cough, and breathlessness *Differential diagnosis*: pneumonia. CXR—shows lung infiltration confined within the treatment volume *Management*: steroids—start with 40mg od prednisolone and reduce over a period of weeks as improvement occurs. ❗ Pulmonary fibrosis may occur >12mo after treatment
Cerebral oedema	Can occur after cranial irradiation. Steroid dose is ↓ after completion of radiotherapy. Consider ↑ dose again
Memory loss	Depending on area irradiated, short- and long-term memory problems can occur after cranial irradiation. Treatment is supportive; some recovery may occur
Somnolence syndrome	Within a few weeks of brain irradiation. *Presents with:* nausea/vomiting; anorexia; dysarthria; ataxia; profound lethargy. Treatment is supportive. Recovery may occur spontaneously

Chapter 28

Palliative care

Palliative care in general practice *1012*
Pain and general debility *1014*
Anorexia, nausea, and vomiting *1016*
Other GI problems *1018*
Skin, neurological, and orthopaedic problems *1020*
Respiratory problems *1022*
Vascular problems *1024*
Problems with mental well-being *1026*
The last 48 hours *1028*
Syringe drivers *1030*

> *'Any man's death diminishes me because I am involved in mankind'*
>
> *Devotions Meditation* 17, John Donne 1572–1631.

Patient/family advice and support

Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk

Palliative care in general practice

❗ Death is the natural end to life—not a failure of medicine

Palliative care starts when the emphasis changes from curing disease and prolonging life to relieving symptoms and maintaining well-being or 'quality of life'. On average, GPs have 1–2 patients with terminal disease at any time and can get more personally involved with them than other patients.

End-of-life care (EOLC) 75% of deaths are 'predictable' and follow a period of chronic illness where end-of-life care (for those likely to die in <12mo) would be appropriate (Figure 28.1).

People are more likely to talk about end of life with their GP than any other professional, but only 33% of GPs are confident to initiate a discussion with a patient about end-of-life issues.

Problems arising are a complex mix of physical, psychological, social, cultural, and spiritual factors involving both patients and their carers. To respond adequately, good lines of communication and close multidisciplinary teamwork is needed. Local palliative care teams are valuable sources of advice and support.

Advanced care planning EOLC is often delivered across health, social care, and voluntary sector services. Planning ahead with the patient can enable the team around them to ensure their needs are met and their wishes communicated should they themselves lose capacity. Details in an advanced care plan may include:

- Preferences around symptom control
- Preferences for care including 'Allow a Natural Death' directives
- Family members or any legal proxies that the patient would like to be involved in decisions about their care
- Advance decisions to withhold treatment (➔ p. 97)
- Discussion about preferred place of death—60–67% of people would prefer to die at home; currently 53% die in hospital but 40% have no medical necessity to die there
- What the patient would like to happen with their body after death. This would include any cultural or religious practices as well as organ or tissue donation

Preferred priorities for care (PPC) The PPC document is a tool for discussion and recording of EOLC wishes. It is available to download from the NHS EOLC website.

The Gold Standards Framework Aims to improve quality of palliative care provided by the primary care team by improving the practice-based organization of care of dying patients. The Framework focuses on: optimizing continuity of care, teamwork, advanced planning (including OOH), symptom control, and patient, carer, and staff support. Evaluation data show the framework increases the proportion of patients dying in their preferred place and improves quality of care as perceived by the practitioners involved.

One chance to get it right The Leadership Alliance for the Care of Dying People (LACDP) released ‘One chance to get it right’ in 2014. This includes 5 priorities for care of dying patients—but avoids a ‘one-size-fits-all tick box’ approach. These priorities are:

1. Recognition Of the possibility that a person may die within the next few days or hours and communicating that clearly
2. Sensitive communication Between health and social care professionals, the dying person, and family/friends
3. Decision-making Ensuring the dying person and those identified as important to him/her are involved in decisions about treatment and care to the extent the dying person wants
4. Family wishes The needs of families and others identified as important to the dying person are actively explored, respected, and met as far as possible
5. Individual plan of care is agreed which includes food and drink, symptom control, and psychological, social, and spiritual support. This should be reviewed regularly and updated as needed

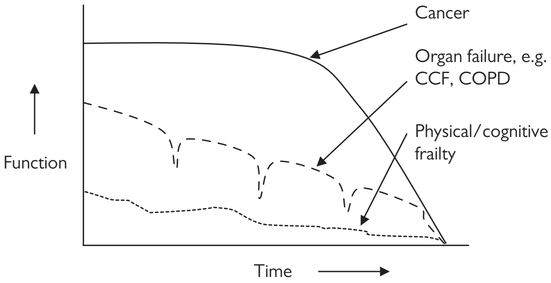

Figure 28.1 Trajectories of decline at the end of life

Further information

Dying Matters 🖱 www.dyingmatters.org
GMC (2010) Treatment and care towards the end of life. 🖱 www.gmc-uk.org/static/documents/content/Treatment_and_care_towards_the_end_of_life_-_English_1015.pdf
Gold Standards Framework 🖱 www.goldstandardsframework.org.uk
Help the Hospices Directory of UK hospice/palliative care services. 🖱 https://www.hospiceuk.org/about-hospice-care/find-a-hospice
LACDP (2014) One chance to get it right. 🖱 https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/323188/One_chance_to_get_it_right.pdf
NHS England End of life care. 🖱 https://www.england.nhs.uk/eolc/
Scottish Government Death and end of life. 🖱 https://www2.gov.scot/Topics/Health/Quality-Improvement-Performance/peolc

Pain and general debility

Pain control This is the cornerstone of palliative care. Cancer pain is multifactorial—be aware of physical and psychological factors.

Principles of pain control ➔ p. 182

Pain-relieving drugs ➔ p. 184

Management of specific types of pain Table 28.1

Weakness, fatigue, and drowsiness Almost a universal symptom.

Reversible causes

- Drugs—opioids, benzodiazepines, steroids (proximal muscle weakness), diuretics (dehydration and biochemical abnormalities), antihypertensives (postural hypotension)
- Emotional problems—depression, anxiety, fear, apathy
- Biochemical abnormalities—hypercalcaemia, DM, electrolyte disturbance, uraemia, liver disease, thyroid dysfunction
- Anaemia
- Infection
- Poor nutrition
- Prolonged bed rest
- Raised intracranial pressure (drowsiness only)

Management Treat reversible causes. Provide advice on modification of lifestyle. If drowsiness/fatigue persists, consider a trial of dexamethasone 4mg/d or an antidepressant. Although steroids make muscle wasting worse, in the short term they may improve general fatigue. Provide psychological support to patients and carers. Consider referral to physiotherapy, review aids and appliances, review home layout (possibly with referral to OT), and/or review home care.

Anaemia Do not check for anaemia if no intention to transfuse.

- If Hb <10 g/dL and symptomatic Treat any reversible cause (e.g. iron deficiency, GI bleeding 2° to NSAIDs). Consider transfusion
- If transfused Record whether any benefit is derived (as if not, further transfusions are futile) and the duration of benefit (if <3 wk—repeat transfusions are impractical). Monitor for return of symptoms; repeat FBC and arrange repeat transfusion as needed

Hypercalcaemia Occurs with 10% malignant tumours—particularly myeloma (>30%) and breast cancer (40%).

> ❗ Always suspect hypercalcaemia if someone is iller than expected for no obvious reason. Untreated hypercalcaemia can be fatal.

Presentation and differential diagnosis ➔ p. 336

Management Depending on the general state of the patient, make a decision whether to treat the hypercalcaemia or not. If a decision is made *not* to treat, provide symptom control and do not check the serum calcium again. Active treatment—➔ p. 336

Patient advice and support

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Table 28.1 Management of specific types of pain

Type of pain	Management
Bone pain (Deep gnawing pain. Worse on moving/weight bearing)	• Try NSAIDs and/or strong opioids • Consider referral for palliative radiotherapy, strontium treatment (prostate cancer), or IV bisphosphonates (↓ pain in myeloma, breast, and prostate cancer) • Refer to orthopaedics if any lytic metastases at risk of fracture, for consideration of pinning
Abdominal visceral pain (Sharp ache. Worse on bending or breathing)	• **Constipation** The most common cause—➔ p. 1019 • **Colic** Try loperamide 2–4mg qds or hyoscine hydrobromide 300 micrograms tds sublingually. Hyoscine butylbromide 20–60mg/24h can also be given via syringe driver • **Liver capsule pain** Dexamethasone 4–8mg/d. Titrate dose to control pain. Alternatively try NSAID + proton pump inhibitor cover • **Gastric distention** Try an antacid ± an anti-foaming agent. Alternatively, a prokinetic may help, e.g. metoclopramide • **Upper GI tumour** Often neuropathic element of pain—coeliac plexus block may help—refer to palliative care • **Consider drug causes** NSAIDs are a common cause • **Acute/subacute obstruction** ➔ p. 1019
Neuropathic pain (Burning/shooting pain associated with altered sensation)	• Usually only partially responsive to opioids—titrate to the maximum tolerated dose • If inadequate, add a neuropathic agent, e.g. amitriptyline 10–25mg nocte—↑ as needed every 2wk to 75–150mg. *Alternatives*: gabapentin, pregabalin, duloxetine, or clonazepam • If pain is due to nerve compression resulting from tumour, dexamethasone 4–8mg od may help • *Other options*: TENS; nerve block; topical lidocaine patches; specialist treatment options, e.g. ketamine
Rectal pain	• Topical drugs, e.g. rectal steroids • Tricyclic antidepressants, e.g. amitriptyline 10–100mg nocte • Anal spasms—glyceryl trinitrate ointment 0.1–0.2% bd • Referral for local radiotherapy
Muscle pain	• Paracetamol and/or NSAIDs • Muscle relaxants, e.g. diazepam 5–10mg od, baclofen 5–10mg tds, dantrolene 25mg od, ↑ weekly to 75mg tds • Physiotherapy, relaxation, heat pads
Bladder pain/spasm	• Treat reversible causes. ↑ fluids. Toilet regularly • Try oxybutynin 5mg tds, tolterodine 2mg bd, propiverine 15mg od/bd/tds, or trospium 20mg bd • Amitriptyline 10–75mg nocte is often effective • If catheterized—try instilling 20mL of intravesical bupivacaine 0.25% for 15min tds or oxybutynin 5mL in 30mL od/bd/tds • NSAIDs can be useful • Steroids, e.g. dexamethasone 4–8mg od may ↓ tumour-related bladder inflammation • In the terminal situation, hyoscine butylbromide 60–120mg/24h or glycopyrronium 0.4–0.8mg/24h sc can help
Pain of short duration	e.g. dressing changes—try a breakthrough dose of oral morphine 20min prior to the procedure.

Anorexia, nausea, and vomiting

Anorexia May improve by treating nausea, mouth problems, pain, and psychological distress. Advise small, appetizing meals frequently in comfortable surroundings.

Drugs that may be helpful

- Alcohol prior to meals
- Metoclopramide prior to meals—to prevent feeling of satiety caused by gastric stasis
- Dexamethasone 2–4mg od or prednisolone 15–30mg od for short-term appetite enhancement

General principles of management of nausea and vomiting

- **Assess** Try to identify likely cause—Table 28.2
- **Review medication** Could medication be the cause? Which antiemetics have been used before and how effective were they?
- **Try non-drug measures**
- **Choose an antiemetic** If cause can be identified, choose an antiemetic appropriate for the cause (Table 28.2). Use the antiemetic ladder (Figure 28.2). Administer antiemetics regularly rather than prn and choose an appropriate route of administration
- **Review frequently** Is the antiemetic effective? Has the underlying cause of the nausea/vomiting resolved? Avoid changing antiemetic before it has been given an adequate trial at maximum dose

! If there is >1 cause for nausea/vomiting you may need >1 drug.

Route of administration

- **For prophylaxis of nausea and vomiting** Use po medication
- **For established nausea or vomiting** Consider a parenteral route, e.g. syringe driver (➔ p. 1030)—persistent nausea may ↓ gastric emptying and drug absorption. Once symptoms are controlled, consider reverting to an oral route

Non-drug measures Do not forget non-drug measures to ↓ nausea:

- Avoidance of food smells and unpleasant odours
- Relaxation/diversion/anxiety management
- Acupressure/acupuncture

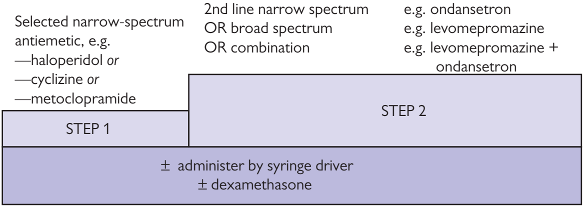
Selected narrow-spectrum antiemetic, e.g.
—haloperidol *or*
—cyclizine *or*
—metoclopramide
2nd line narrow spectrum
OR broad spectrum
OR combination
e.g. ondansetron
e.g. levomepromazine
e.g. levomepromazine + ondansetron
STEP 1
STEP 2
± administer by syringe driver
± dexamethasone

Figure 28.2 The antiemetic ladder

Table 28.2 Causes of vomiting and choice of antiemetic

Mechanism of vomiting	Antiemetic
Drug/toxin induced or metabolic (e.g. hypercalcaemia)	• Haloperidol (1.5–5mg nocte) • Levomepromazine (5mg stat or 6.25mg nocte) • If persistent nausea due to opioids, consider changing opioid
Chemotherapy/ radiotherapy	• Granisetron (1mg bd) or ondansetron (8mg bd PO or 16mg od PR)—chemo- or radiotherapy-induced vomiting • Haloperidol 1.5–5mg nocte—radiotherapy-induced vomiting • Dexamethasone 4–8mg daily PO/sc—often given as part of a chemotherapy regimen • Metoclopramide 20mg tds
↑ *intracranial pressure*	• Dexamethasone 4–16mg/d • Cyclizine 50mg bd/tds (or 150mg/d via syringe driver)
Anxiety, fear, or pain	• Benzodiazepines, e.g. diazepam 2–10mg/d or midazolam sc • Cyclizine 50mg bd/tds • Levomepromazine 6–25mg/d
Motion/position	• Cyclizine 50mg tds PO/sc/IM • Hyoscine PO (300 micrograms tds) or transdermally (1mg/72h) • Prochlorperazine PO (5mg qds) or buccal (3–6mg bd)
Gastric stasis[a]	Metoclopramide 10mg tds (particularly if multifactorial with gastric stasis and a central component)
Gastric irritation	• Stop the irritant if possible, e.g. stop NSAIDs • Proton pump inhibitors, e.g. lansoprazole 30mg od or omeprazole 20mg od • Antacids • Misoprostol 200 micrograms bd—if caused by NSAIDs
Constipation	Laxatives/suppositories/enemas
Intestinal obstruction	• Refer for surgery if appropriate • Cyclizine, haloperidol, or levomepromazine • Dexamethasone 4–8mg/d—antiemetic and ↓ obstruction • If vomiting cannot be controlled consider referral for venting gastrostomy or antisecretory agents (e.g. octreotide)
Cough induced	➔ p. 1022
Unknown cause	• Cyclizine 50mg tds or 150mg/d via syringe driver • Levomepromazine 6–25mg/d • Dexamethasone 4–8mg daily PO/ sc • Metoclopramide 10–20mg tds/qds PO

[a] Vomiting of undigested food without nausea soon after eating.

❗ Drugs with antimuscarinic effects (e.g. cyclizine) antagonize prokinetic drugs (e.g. metoclopramide)—where possible, do not use concurrently.

Other GI problems

Mouth problems Review medication making the mouth sore/dry. Refer to the DN for mouth care advice (e.g. use a toothbrush to keep the tongue clean). Consider mouthwashes for pain. *Specific measures*:

- Oral thrush—treat with fluconazole 50mg od for 7d and soak dentures in Milton® fluid for ≥12h to prevent reinfection.
- Painful mouth—benzydamine mouthwash ± lidocaine® spray
- Ulcers—hydrocortisone pellets topically qds after eating and nocte
- Oral cancer pain—topical NSAIDs, e.g. soluble aspirin or diclofenac
- Chemotherapy-induced ulcers—sucralfate suspension
- Dry mouth—review medication that might be causing dry mouth, e.g. antidepressants, opioids. Try salivary stimulants, e.g. iced water, pineapple chunks, chewing gum, boiled sweets or mints. Consider saliva substitutes, e.g. Glandosane® spray
- Radiotherapy-induced dryness—pilocarpine
- Excessive salivation—amitriptyline 10–100mg nocte, hyoscine or glycopyrronium via syringe driver

Dysphagia May be due to physical obstruction by tumour or functional obstruction (neurological deficit).

- Treat the cause if possible, e.g. celestin tube
- If the patient is hungry and wishes to be fed, consider referral for a percutaneous endoscopic gastrostomy (PEG)
- If the patient does not wish to have a PEG, ask whether s/he would like sc fluids and treat symptomatically with mouth care, anxiolytics, analgesia, and sedation

Hiccup A distressing symptom. Treatment is often unsatisfactory.

- General measures Rebreathing with a paper bag; pharyngeal stimulation with cold water or a teaspoon of granulated sugar
- Peripheral hiccups Irritation of the phrenic nerve/diaphragm—try metoclopramide (10mg tds), antacids containing simeticone, dexamethasone (4–12mg/d), or ranitidine (150mg bd)
- Central hiccups Due to medullary stimulation, e.g. ↑ ICP, uraemia—try chlorpromazine (10–25mg tds/qds), dexamethasone (4–12mg/d), nifedipine (10mg tds), or baclofen (5mg bd)

Ascites Free fluid in the peritoneal cavity. Common with ovarian cancer (50%). Presents with abdominal distention, shifting dullness to percussion ± fluid thrill. Depending on clinical state consider referring for radio- or chemotherapy if appropriate. *Symptom control*:

- Give analgesia for discomfort and try prokinetics, e.g. domperidone or metoclopramide 10mg tds for 'squashed stomach syndrome'
- Consider referral for paracentesis and/or peritoneovenous shunt
- Try diuretics—furosemide 20–40mg od and/or spironolactone 100–600mg od. May take a week to produce maximal effect. ❗ Monitor albumin level—if low, diuretics make ascites worse
- Dexamethasone 2–4mg daily may help—discontinue if not effective

Gut fistula Connection from the gut to another organ—commonly skin, bladder, or vagina. Bowel fistulae are characterized by air passing through the fistula channel. If well enough for surgery, refer. If not fit for surgery, consider referring to palliative care for octreotide.

Bowel obstruction Causes nausea and vomiting, abdominal distension, and altered bowel habit. *Signs*: haemodynamic instability, tender distended abdomen, and ↓ or absent bowel sounds. Offer stents to appropriate patients but medical management in the community in those undergoing palliation may be the best course of action:

- **Subacute/functional obstruction** Metoclopramide 30–60mg/24h via syringe driver (stop if complete bowel obstruction)
- **Complete obstruction** If the patient is not admitted try cyclizine then haloperidol, then levomepromazine via syringe driver. Seek specialist advice if treatment resistant

Constipation Passage of hard stools less frequently than the patient's norm. Very common symptom. Occult presentations may include:

- Confusion
- Urinary retention
- Abdominal pain
- Overflow diarrhoea
- Loss of appetite
- Nausea/vomiting

❗ Constipation can herald spinal cord compression (➔ p. 453). If suspected, do a full neurological examination.

Management Pre-empt constipation by putting everyone at risk (e.g. patients on opioids) on regular aperients. ↑ fluid intake.

- Treat reversible causes (e.g. alter diet by adding fibre)
- Treat with regular stool softener (e.g. lactulose, macrogol) ± regular bowel stimulant (e.g. senna) or a combination drug (e.g. co-danthrusate). Titrate dose against response
- If that is ineffective, consider adding rectal measures. If soft stools and lax rectum—try bisacodyl suppositories (❗ must come into direct contact with rectum); if hard stools—try glycerol suppositories—insert into the faeces and allow to dissolve
- If still not cleared refer to the DN for lubricant ± stimulant enema
- Once cleared leave on a regular aperient with instructions to ↑ aperients if constipation recurs

Diarrhoea Clarify what the patient/carer means by diarrhoea. Less common than constipation but can be distressing for patients and difficult for carers—especially if incontinence results. *Management*:

- ↑ fluid intake—small amounts of clear fluids frequently
- Screen for infection (including pseudomembranous colitis if diarrhoea after a course of antibiotics) and treat if necessary
- Ensure no overflow diarrhoea 2° to constipation; no excessive/erratic laxative use; and no other medication is causing diarrhoea
- Radiation-induced diarrhoea—consider ondansetron 4mg tds or aspirin 300–600mg tds (↓ intestinal electrolyte and water secretion caused by prostaglandins)
- Consider pancreatic enzyme supplements, e.g. Creon® 25000 tds prior to meals if fat malabsorption (e.g. 2° to pancreatic carcinoma)
- Otherwise treat symptomatically with codeine phosphate 30–60mg qds or loperamide 2mg tds/qds
- Refer to palliative care if unable to control symptoms

Skin, neurological, and orthopaedic problems

Bed sores Due to pressure necrosis of the skin. Immobile patients are at high risk—especially if frail ± incontinent. Likely sites of pressure damage—shoulder blades, elbows, spine, buttocks, knees, ankles, and heels. Bed sores heal slowly in terminally ill patients and are a source of discomfort and stress for both patients and carers (who often feel guilty that a pressure sore is a mark of poor care).

- If at risk refer to the DN or palliative care nursing team for advice on prevention of bed sores—protective mattresses and cushions, incontinence advice, advice on positioning and movement
- Warn carers to make contact with the DN or palliative care nursing team if a red patch does not improve 24h after relieving the pressure on the area
- Treat any sores that develop aggressively; admit if not resolving

Wound care Large wounds can have major impact on quality of life. Patients with advanced disease have major risk factors for development and poor healing of wounds—immobility, poor nutrition, skin infiltration ± breakdown due to malignancy. Skin infiltration causing ulceration or fungating wounds can be particularly distressing.

Management The primary aim is comfort. Healing is a 2° aim and may be impossible. Always involve the DN and/or specialist palliative care nursing team early. Many hospitals also have wound care specialist nurses who are valuable sources of advice.

Specific management problems Table 28.3

Raised intracranial pressure Occurs with 1° or 2° brain tumours. Characterized by:

- Headache—worse on lying
- Vomiting
- Confusion
- Diplopia
- Convulsions
- Papilloedema

Management

- Unless a terminal event, refer urgently to neurosurgery for assessment. Options include insertion of a shunt or cranial radiotherapy
- If no further active treatment is appropriate, start symptomatic treatment—raise the head of the bed, start dexamethasone 16mg/d (stop if no response in 1wk), analgesia

Spinal cord compression ➔ p. 453

Bone fractures Common in advanced cancer due to osteoporosis, trauma as a result of falls, or metastases. Have a low index of suspicion if a new bony pain develops. Treat with analgesia. Unless in a very terminal state, confirm the fracture on X-ray and refer to orthopaedics or radiotherapy urgently for consideration of fixation (long bones, wrist, neck of femur) and/or radiotherapy (rib fractures, vertebral fractures).

Fracture of a long bone can present as acute delirium in the elderly.

Table 28.3 Common wound management problems

Problem	Management
Pain	Exclude infection; ensure the dressing is comfortable; limit frequency of dressing changes Ensure adequate background analgesia; consider additional analgesia for dressing changes and/or topical opioids on the dressing
Excessive exudate	Use high-absorbency dressings with further packing on top ± plastic pads to protect clothing Change the top layer of the dressing as often as needed but avoid frequent changes of the dressing placed directly on the wound Protect the surrounding skin with a barrier cream/spray
Necrotic tissue	Use desloughing agents Referral for surgical debridement may be necessary
Bleeding	Prevent bleeding during dressing changes by: • Avoiding frequent dressing changes • Using non-adherent dressings or dressings which liquefy and can be washed off (e.g. alginate dressings) and • Irrigating the wound with saline to remove dressings If there is surface bleeding—put pressure on the wound; if pressure is not working try: • Kaltostat® • Adrenaline—1mg/mL (or 1:1000) on a gauze pad, or • Sucralfate liquid—place on a non-adherent dressing and apply firmly to the bleeding area Consider referral for radiotherapy or palliative surgery (e.g. cautery)
Odour	Treat with systemic and/or topical metronidazole Charcoal dressings can be helpful Seal the wound, e.g. with an additional layer of cling film dressing Try disguising the smell with deodorizers (e.g. Nilodor®) used sparingly on top of the dressing—short-term measure. Long term, the deodorant smell often becomes associated with the smell of the wound for the patient
Infection	Usually chronic and localized Irrigate the wound with warm saline or under running water in the shower/bath If the surrounding skin is inflamed—swab the wound and send for M,C&S then start oral antibiotics, e.g. flucloxacillin 250–500mg qds or erythromycin 250–500mg qds. Alter antibiotics depending on sensitivities of the organisms grown

Respiratory problems

Cough Troublesome symptom. Prolonged bouts of coughing are exhausting and frightening—especially if associated with breathlessness and/or haemoptysis.

Haemoptysis ➜ p. 267

Breathlessness Affects 70% of terminally ill patients. It is usually multifactorial. Breathlessness always has a psychological element—being short of breath is frightening. *Causes*: Figure 28.3.

Management of cough and breathlessness

General non-drug measures

- Generally reassure. Explain reasons for breathlessness/cough and adaptations to lifestyle that might help, e.g. sitting up straight
- Breathing exercises can help—refer to physiotherapy
- Exclude treatable causes (Box 28.1 and Figure 28.3)
- Steam inhalations/nebulized saline help with tenacious secretions
- Try a stream of air over the face if the patient is breathless, e.g. fan, open window

General drug measures

- Oral or sc opioids ↓ the subjective sensation of breathlessness and may alleviate cough, e.g. start with 2.5mL of 10mg/5mL morphine sulfate solution PO every 4h and titrate upwards. If already taking opioids, ↑ dose by 25%. Titrate dose until symptoms are controlled or the patient experiences side effects
- Try benzodiazepines—2–5mg diazepam od/bd for associated anxiety + lorazepam 1–2mg sublingually prn in between. Diazepam also acts as a central cough suppressant—try 2–10mg tds for cough
- Hyoscine 400–600 micrograms 4–8-hourly (or 0.6–2.4mg/24h via syringe driver) and/or inhaled or nebulized ipratropium ↓ secretions

Specific measures

- **Chest infection** Treat with nebulized saline to make secretions less viscous ± antibiotics (if not considered a terminal event)
- **Post-nasal drip** Steam inhalations, steroid nasal spray or drops ± antibiotics
- **Laryngeal irritation** Try inhaled steroids, e.g. Clenil® 100 micrograms/actuation 2 puffs bd
- **Bronchospasm** Try bronchodilators ± inhaled or oral steroids. ❗ salbutamol may help cough even in the absence of wheeze
- **Gastric reflux** Try antacids containing simeticone
- **Lung cancer** Try inhaled sodium cromoglicate 10mg qds; local anaesthesia using nebulized bupivacaine or lidocaine can be helpful—refer for specialist advice (avoid eating/drinking for 1h afterwards to avoid aspiration). Palliative radiotherapy or chemotherapy can also relieve cough in patients with lung cancer—refer

Stridor Coarse wheezing sound that results from the obstruction of a major airway, e.g. larynx.

Management

- Corticosteroids (e.g. dexamethasone 16mg/d) can give relief
- Consider referral for radiotherapy or endoscopic insertion of a stent if appropriate
- If a terminal event—sedate with high doses of midazolam (10–40mg repeated as needed)

Box 28.1 Reversible causes of cough

- Infection
- Bronchospasm
- Gastro-oesophageal reflux
- Aspiration
- Drug induced, e.g. ACE inhibitors
- Treatment related, e.g. total body irradiation
- Malignant bronchial obstruction/lung metastases
- Heart failure
- Secretions
- Pharyngeal candidiasis

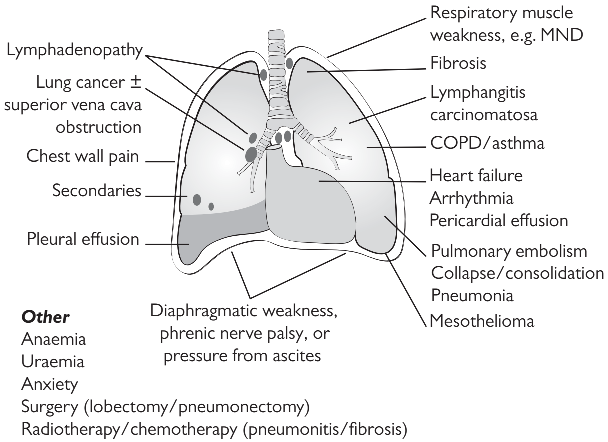
Lymphadenopathy
Lung cancer ± superior vena cava obstruction
Chest wall pain
Secondaries
Pleural effusion
Respiratory muscle weakness, e.g. MND
Fibrosis
Lymphangitis carcinomatosa
COPD/asthma
Heart failure
Arrhythmia
Pericardial effusion
Pulmonary embolism
Collapse/consolidation
Pneumonia
Mesothelioma
Diaphragmatic weakness, phrenic nerve palsy, or pressure from ascites
Other
Anaemia
Uraemia
Anxiety
Surgery (lobectomy/pneumonectomy)
Radiotherapy/chemotherapy (pneumonitis/fibrosis)

Figure 28.3 Causes of breathlessness

Patient/family advice and support

Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk

Vascular problems

Bleeding/haemorrhage In all patients likely to bleed (e.g. in end-stage leukaemia), pre-warn carers and give them a strategy.

Severe, life-threatening bleed Make a decision whether the cause of the bleed is treatable or a terminal event. This is best done in advance but bleeding cannot always be predicted.

- Severe bleed—active treatment ➜ p. 1062
- Severe bleed—no active treatment:
  - Stay with the patient
  - Give sedative medication, e.g. midazolam 20–40mg sc or IV, or lorazepam 1–2mg sublingually. If in pain, consider sc opioid
  - Support carers as big bleeds are extremely distressing

Non-life-threatening bleed: first aid measures

- In all cases—reassure; monitor frequently
- Surface bleeding—pressure on wound; if pressure is not working try Kaltostat® or adrenaline (1mg/mL or 1:1000) on a gauze pad
- Nose bleeds—nasal packing or cautery

Non-life-threatening bleed: follow-up treatment Follow-up is directed at cause if appropriate:

- Anticoagulation—consider stopping; if coumarin anticoagulant—check INR
- Treat infection that might exacerbate a bleed
- Consider ↓ bleeding tendency with tranexamic acid 500mg qds
- Upper GI bleeding—stop NSAIDs, start proton pump inhibitor in double standard dose and consider referral for gastroscopy
- Lower GI bleeding—consider rectal steroids to ↓ inflammation or oral tranexamic acid ± referral for colonoscopy
- Radiotherapy—consider referral if haemoptysis, cutaneous bleeding, or haematuria
- Referral for chemotherapy or palliative surgery (e.g. cautery) are also options

Anaemia ➜ p. 1014

Superior vena cava (SVC) obstruction Due to infiltration of the vessel wall, clot within the SVC, or extrinsic pressure. 75% are due to 1° lung cancer (3% of patients with lung cancer have SVC obstruction). Lymphoma and clotting associated with long central lines are the two other major causes.

Presentation

- Shortness of breath/stridor
- Headache worse on stooping ± visual disturbances ± dizziness and collapse
- Swelling of the face—particularly around the eyes, neck, hands, and arms, and/or injected cornea
- *Examination*: look for non-pulsatile distention of neck veins and dilated collateral veins (seen as small dilated veins over the anterior chest wall below the clavicles) in which blood courses downwards

Management

- Treat breathlessness (2.5–5mL of 10mg/5mL morphine sulfate solution every 4h ± benzodiazepine depending on level of anxiety)
- Start corticosteroid (dexamethasone 16mg/d)
- Refer urgently for oncology opinion. Palliative radiotherapy has a response rate of 70%. Stenting ± thrombolysis is also an option

Lymphoedema Due to obstruction of lymphatic drainage resulting in oedema with high protein content. Affects ≥1 limbs ± adjacent trunk. If left untreated, lymphoedema becomes increasingly resistant to treatment due to chronic inflammation and subcutaneous fibrosis. Cellulitis causes rapid ↑ in swelling. *Causes*:

- Axillary, groin, or intrapelvic tumour
- Axillary or groin surgery (including biopsy)
- Postoperative infection/radiotherapy

Presentation

- Swollen limb ± pitting
- Impaired limb mobility and function
- Discomfort/pain related to tissue swelling and/or shoulder strain
- Neuralgia pain—especially when axillary nodes are involved
- Psychological distress.

Management Table 28.4

Table 28.4 Management of lymphoedema

Avoid injury to limb	In at-risk patients (e.g. patients who have had breast cancer with axillary clearance) or those with lymphoedema, injury to the limb may precipitate or worsen lymphoedema. Do not take blood from the limb or use it for IV access or vaccination
Skin hygiene	Skin care with emollients Topical treatment of fungal infection Systemic treatment of bacterial infection, e.g. clarithromycin 500mg bd for 14d
External support	Intensive—with compression bandages Maintenance—with lymphoedema sleeve (contact breast care specialist nurse for more information on obtaining sleeves)
Exercise	Gentle daily exercise of affected limb gradually ↑ range of movement ❗ Must wear a sleeve/bandages when doing exercises
Massage	Very gentle finger tip massage in the line of drainage of lymphatics
Diuretics	If the condition has developed or deteriorated since prescription of corticosteroid or NSAID, or if there is a venous component, consider a trial of diuretics Otherwise diuretics are of no benefit

Patient/family advice and support

Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk

Problems with mental well-being

Anxiety All patients with terminal disease are anxious at times for a variety of reasons including fear of uncontrolled symptoms, of being left alone to die, and spiritual angst. When anxiety starts interfering with quality of life, intervention is justified.

Management: non-drug measures Often all that is needed:

- Acknowledgement of the patient's anxiety
- Full explanation of questions + written information as needed
- Support—self-help/patient groups, day care, specialist home nurses (e.g. Macmillan Nurses), religious groups as appropriate
- Relaxation training and training in breathing control
- Physical therapies, e.g. aromatherapy, art therapy, exercise

Management: drug measures

- **Acute anxiety** Try lorazepam 1–2mg sublingually or diazepam 2–10mg prn
- **Chronic anxiety** Try an antidepressant, e.g. fluoxetine 20mg od. Alternatives include regular diazepam, e.g. 5–10mg od/bd, haloperidol 1–3mg bd/tds, or β-blockers, e.g. propranolol 40mg od–tds—watch for postural hypotension

If anxiety is not responding to simple measures, seek specialist help from either the psychiatric or palliative care team.

Depression A terminal diagnosis commonly makes patients sad. 10–20% of terminally ill patients develop clinical depression but, in practice, it is often difficult to decide whether a patient is depressed or just appropriately sad about his/her diagnosis and its implications. Many symptoms of terminal disease (e.g. poor appetite) are also symptoms of depression so screening questionnaires for depression are often unhelpful. If in doubt, a trial of antidepressants can help.

Assessment of suicide risk Ask about suicidal ideas and plans in a sensitive but probing way. It is a common misconception that asking about suicide can plant the idea into a patient's head and make suicide more likely. Evidence is to the contrary.

Management: non-drug measures

- Support—e.g. day and/or respite care; carers group; specialist nurse support (e.g. Macmillan Nurse; CPN); ↑ help in the home
- Relaxation and/or simple mindfulness techniques—often ↑ the patient's feeling of control over the situation
- Explanation—of worries/problems/concerns about the future
- Physical activity—exercise; writing

Management: drug measures

- Consider starting an antidepressant—➔ p. 982
- All antidepressants take ~2wk to work
- If immediate effect is required consider using flupentixol 1mg od (beware as can cause psychomotor agitation)

❗ If not responding or suicidal, refer for psychiatric opinion.

Terminal anguish and spiritual distress Characterized by overwhelming distress. Often related to unresolved conflict, guilt, fears, or loss of control.

Anxiety can be increased if

- Patients are unaware of the diagnosis, but feel that people are lying to them
- The patient is breathless, haemorrhaging, or has constant nausea or diarrhoea
- Weak religious conviction—convinced believers and convinced non-believers have less anxiety
- There are young dependent children or other dependent relatives
- Patients have unfinished business to attend to, such as legal affairs

Action Listening can itself be therapeutic. Talk to the patient, if possible, about dying and try to break down fears into component parts. Address fears that can be dealt with. As a last resort, and after discussion with the patient (where possible) and/or relatives, consider sedation.

Confusion (acute delirium) ➔ p. 988

Insomnia ➔ p. 168

Patient/family advice and support

Macmillan Cancer Support ☎ 0808 808 0000 🖱 www.macmillan.org.uk

The last 48 hours

It is notoriously difficult to predict when death will occur. Symptoms and signs of death approaching include:

- Day-by-day deterioration
- Gaunt appearance
- Difficulty swallowing medicines
- ↓ intake of food and fluids
- Profound weakness—needs assistance with all care, may be bedbound
- Drowsy or ↓ cognition—often unable to cooperate with carers

Patients' wishes Dying is a unique and special event for each individual. Helping to explore a patient's wishes about death and dying should not be a discussion left to the last 24h.

Different cultures Different religious and cultural groups have different approaches to the dying process. Be sensitive to cultural and religious beliefs. Never assume; if in doubt ask a family member or your local palliative care team.

Advance directives/lasting power of attorney ➔ p. 96, p. 97

Goals of treatment in the last 48h Good nursing care is the mainstay of treatment in the last 48h

- Ensure patients are comfortable—physically, emotionally, and spiritually. Try to make the end of life peaceful/dignified and in line with the wishes expressed during the advanced care planning stage
- Support patients and carers so that the experience of death for those left behind is as positive as it can be

The role of the GP

- Ensure new problems do not develop, e.g. use of appropriate mattresses and measures to prevent bed sores
- Stop all unnecessary medication
- Treat specific symptoms, e.g. dry mouth
- Think ahead—discuss possible future treatment options, e.g. syringe driver, buccal, PR, or transcutaneous preparations if the oral route is no longer available; use of strong analgesia with sedative effect
- Ensure there is a clearly agreed management plan that the patient/family are happy with. Anticipate likely needs of the patient, e.g. define clearly what to do in the event of a symptom arising/worsening; ensure drugs + equipment that may be needed are in the home; inform the OOH service.

Out-of-hours providers Alert OOH providers (primary care, district nursing, ambulance service) if a patient is dying at home to ensure appropriate response and avoid unnecessary admission. Arrange anticipatory medication containing drugs that might be needed to be at the patient's home for use if needed outside working hours.

Assessment of patient needs Ask which problems are causing the patient/carers most concern and address those concerns where possible. Patients often under-report symptoms.

Physical examination Keep examination to a minimum to avoid unnecessary interference. Check sites of discomfort/pain suggested by history or non-verbal cues, e.g. mouth, bladder, and bowel.

Psychological assessment Find out what the patient wants to know. Gently assessing how patients feel about their disease and situation can shed light on their needs and distress.

Investigations Any investigation at the end of life should have a clear and justifiable purpose (e.g. excluding a reversible condition where treatment would make the patient more comfortable). The need for investigations in the terminal stage of illness is minimal.

Excessive respiratory secretion (death rattle) Noisy, moist breathing. Can be distressing for relatives. Reassure that the patient is not suffering or choking. Try repositioning and/or tipping the bed head down (if possible) to ↓ noise. Treat prophylactically—it is easier to prevent than remove accumulated secretions. *Suitable drugs*:

- Glycopyrronium Non-sedative—give 200 micrograms sc stat and review after 1h. If effective, give 200 micrograms every 4h sc or 0.6–1.2mg/24h via syringe driver
- Hyoscine hydrobromide Sedative in high doses—give 400 micrograms sc stat and review response after 30min. If effective, give 400–600 micrograms 4–8-hourly or 0.6–2.4mg/24h via syringe driver. If the patient is conscious and respiratory secretions are not too distressing, it may be more appropriate to use a transdermal patch (Scopaderm® 1.5mg over 3d) or sublingual tablets (Kwells®). Dry mouth is a side effect

Terminal breathlessness Distressing symptom for patients/carers. Support carers in attendance and explain management:

- Diamorphine or morphine Dose depends on whether the patient is already taking an opioid. If taking oral morphine, switch to equivalent sc dosage (Table 8.2, ➔ p. 187). If no previous opioid, start diamorphine 5mg/24h or morphine 10mg/24h via syringe driver. ↑ dose slowly as needed
- Midazolam 5–10mg/24h via syringe driver
- If sticky secretions Try nebulized saline ± physiotherapy

Terminal restlessness *Causes*:

- Pain/discomfort Urinary retention, constipation, pain which the patient cannot tell you about, excess secretions in throat
- Opioid toxicity Causes myoclonic jerking. Dose of morphine may need to be ↓ if a patient becomes uraemic
- Biochemical causes ↑ Ca^{2+}, uraemia—❗ if there is no intention to treat, do not check
- Psychological/spiritual distress

Management Treat reversible causes, e.g. catheterization for retention, hyoscine to dry up secretions. If still restless, treat with a sedative. This does not shorten life but makes the patient/relatives more comfortable. *Suitable drugs*:

Oral:	Via syringe driver:
• Haloperidol 1–3mg tds	• Midazolam 10–100mg/24h or 5mg stat
• Chlorpromazine 25–50mg tds	• Levomepromazine 50–150mg/24h or 6.25mg stat
• Diazepam 2–10mg tds	

Terminal anguish and spiritual distress ➔ p. 1027

Syringe drivers

Syringe drivers are used to aid drug delivery when the oral route is no longer feasible. Indications include:

- Intractable vomiting
- Severe dysphagia
- Patient too weak to swallow
- ↓ conscious level
- Poor gut absorption (rare)
- Poor patient compliance

Types of syringe driver In recent years, many PCOs, hospitals, and hospices have been changing their syringe drivers from the traditionally used blue or green Graseby drivers, which are being phased out. Newer devices with additional safety and monitoring features (e.g. McKinley T34 or Alaris) are now in common use. It is important to find out which devices are used in your locality and how they work.

> ❗ Incorrect use of syringe drivers is a common cause of drug errors. Each PCO should use just one type of syringe driver to ↓ risks of errors.

Drugs that can be used in syringe drivers Table 28.5

General principles Draw up the prescribed 24h medication. The diluent of choice in most cases is water for injection but 0.9% sodium chloride should be used if using levomepromazine, diclofenac, octreotide, or ondansetron; cyclizine should not be diluted with saline. Then set the rate on the syringe driver

❗ Local policies may differ. Hands-on training is essential.

Mixing drugs in syringe drivers Provided there is evidence of compatibility, drugs can be mixed in syringe drivers. Diamorphine and morphine can be mixed with:

- Cyclizine
- Hyoscine hydrobromide
- Hyoscine butylbromide
- Midazolam
- Ondansetron
- Dexamethasone (<4mg/24h)
- Levomepromazine
- Haloperidol
- Metoclopramide
- Glycopyrronium

If combining 2 or 3 drugs in a syringe driver, a larger volume of diluent may be needed (e.g. 20 or 30mL syringe). If >3 drugs are needed in one syringe driver—reassess treatment aims.

Common problems with syringe drivers

- **If the syringe driver runs too slowly** Check it is switched on; check the battery; check the cannula is not blocked
- **If the syringe driver runs too quickly** Check the rate setting
- **Injection site reaction** If there is pain or inflammation, change the injection site

Further information

Sdrivers Syringe driver drug compatibility database 🖱 www.pallcare.info
Palliative Care Adult Network Guidelines 🖱 http://book.pallcare.info/

Table 28.5 Drugs that can be used in syringe drivers

Indication	Drugs
Nausea and vomiting	Haloperidol 2.5–10mg/24h Levomepromazine 5–200mg/24h (causes sedation in 50%) Cyclizine 150mg/24h (may precipitate if mixed with other drugs) Metoclopramide 30–100mg/24h Octreotide 300–600 micrograms/24h (consultant supervision) Hyoscine hydrobromide 20–60mg/24h
Respiratory secretions	Hyoscine hydrobromide 0.6–2.4mg/24h Glycopyrronium 0.6–1.2mg/24h
Restlessness and confusion	Haloperidol 5–15mg/24h Levomepromazine 50–200mg/24h Midazolam 20–100mg/24h (and fitting)
Pain control	Diamorphine ⅓–½ dose of oral morphine/24h Morphine ½–⅔/$_3$ dose of oral morphine/24h Oxycodone ½ dose of oral oxycodone/24h

❗ Subcutaneous infusion solution should be monitored regularly both to check for precipitation (and discoloration) and to ensure the infusion is running at the correct rate.

Chapter 29

Emergencies

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Miscellaneous and environmental emergencies *1108*

Emergency patient encounters

The doctor's bag ➲ p. 68

Emergency calls Nearly all requests for emergency care in the community are made by telephone. *General rules*:

- Train staff to handle distressed callers, recognize serious problems, and act appropriately when such calls are received
- Ensure that it is easy for patients to seek help in an emergency, e.g. by ensuring messages on phone systems are easily heard and contain simple, clear instructions. Worried patients find it difficult to cope with complicated telephone referral systems or messages
- Appear helpful. Keep calm and friendly—even in the event of provocation. Worried callers may appear abrupt or demanding
- Record the time of the call, date, patient's name, address, and a contact telephone number, brief details of the problem, and action taken (even if calls are being recorded)
- Collect only information you need to decide what action is necessary. If the patient needs to be seen, collect enough information to decide where and how quickly the patient should be seen, and whether extra equipment or help is needed
- If giving advice make it simple and in language the patient can understand. Repeat to make sure it has been understood. Consider asking the patient/carer to repeat what you have told them. Always tell callers to ring back if symptoms change or they have further worries
- If a visit is indicated, ensure the address is right and ask for directions if you are not sure where to go. Try to give a rough arrival time
- In some cases (e.g. major trauma, large GI bleeds, suspected MI, burns, overdoses) call for an emergency ambulance at once
- If a call seems inappropriate, consider the reason for it—e.g. depression might provoke recurrent calls for minor ailments

⚠ If in doubt—arrange for the patient to be seen.

Emergency home visits

- Try to stick to the problem you have been called about
- Take a concise history and examine as appropriate
- Make a decision on management and explain it to the patient and any carers in clear and concise terms that they can understand. Repeat advice several times ± write it down
- Record history, examination, management suggested, and advice given for the patient's notes
- Always invite the patient and carers to seek medical help again should symptoms change, the situation deteriorate, or further worries appear
- For inappropriate calls, take time to educate the patient and/or carers about self-management and use of emergency GP visiting services
- Always consider hidden reasons for seemingly unnecessary visits

Being prepared

- Ensure that you have a reliable vehicle with fuel
- Have a street map and an electronic in-car navigation system
- Carry a large, strong torch in the vehicle

- Carry a mobile telephone to enable you to call for help as needed
- Check your drug box is fully stocked and all items are in date
- Check all equipment carried is operational, and carry spare batteries/consumables (e.g. ear pieces for electronic thermometers)
- Carry a list of emergency telephone numbers
- Know which pharmacies have extended opening

Safety and security

- Ensure that you have adequate supplies of personal protective equipment (PPE)
- If performing home visits alone, ensure someone else knows where you are going, when to expect you back, and what to do if you do not return on time
- If going to a call you are worried about, either take someone with you or call the police/ambulance to meet you there before going in
- If you reach a call and find you are uncomfortable, make sure you can get out. Note the layout of the property and make sure you have a clear route to the door
- Set up your mobile phone to call the police or your base at a single touch of a button. Consider carrying an attack alarm
- Have separate bags for drugs and consultation equipment; leave the drug box locked out of sight within your vehicle when doing a visit

Handing over Box 29.1. When passing care to other teams (e.g. emergency services, hospital-at-home services, on-call hospital teams), it is important to provide a clear, concise handover including all necessary information. Good handovers save time and ↓ errors.

Referral letters ➔ p. 66

Assessing severity using the NEWS/NEWS2 score ➔ p. 1036

Box 29.1 SBAR communication tool

Situation

- Who you are—name, what you do, who you work for
- Patient's name, date of birth
- Presenting complaint
- Standard observations—level of consciousness, temperature, pulse, BP, respiratory rate, peripheral oxygen saturation (if available), capillary blood glucose (if relevant), peak flow rate (if relevant)
- Any additional relevant examination findings ± NEWS/NEWS2 score

Background

- Normal state
- Past medical history
- Medication history
- Smoking, alcohol, illicit drug use
- Social circumstances, e.g. home situation, family, carers

Assessment

- Your concerns/worries, e.g. diagnoses to exclude, rapid change in clinical state, possible dangers
- Provisional diagnosis
- Actions already taken, e.g. drugs given (doses, time, and route)

Recommendation (or requests)

- Actions that you would like the receiving team to take
- Anything that the receiving team would like you to do?

Warning signs for acute severe illness

The National Early Warning Score (NEWS) was developed in 2012 to improve detection of and response to clinical deterioration in patients with acute illness. It is widely used both in the UK and other countries across both primary and secondary care. An updated version (NEWS2) was released in 2017.

⚠ The National Early Warning Score should not be used for assessment of patients aged <16y or pregnant women.

Physiological parameters Figure 29.1. The NEWS2 score is based on 6 physiological parameters. A score is allocated to each parameter; the more a parameter varies from the norm, the higher the score. An overall score is generated by adding together scores for each parameter.

Respiratory rate Number of breaths over a minute.

- ↑ respiratory rate is a powerful sign of acute illness
- ↓ respiratory rate may indicate CNS depression or narcosis

Peripheral oxygen saturation Using a pulse oximeter.

- Use 'O_2 saturation 1' for most patients—↑ score by 2 points for patients requiring supplemental oxygen to maintain O_2 saturations
- Use 'O_2 saturation 2' if hypercapnic respiratory failure (e.g. COPD) where normal peripheral oxygen saturation is 88–92%; when given supplementary oxygen, high oxygen saturations in this group can lead to CO_2 retention and clinical deterioration

Systolic BP

- ↓ or falling systolic BP Is most significant in the context of assessing acute illness severity
- Very ↑ systolic BP Could indicate accelerated hypertension
- Diastolic BP Does not add additional information in this context and is not included in the NEWS2, but should be measured and recorded

Pulse rate

- Tachycardia May be indicative of:
  - Circulatory compromise due to sepsis or volume depletion
  - Cardiac failure
  - Pyrexia
  - Pain
  - General distress
  - Cardiac arrhythmia
  - Metabolic disturbance, e.g. hyperthyroidism
  - Drug intoxication, e.g. sympathomimetics or anticholinergic drugs
- Bradycardia is also an important physiological indicator. It may be normal with physical conditioning or due to:
  - Hypothyroidism
  - Hypothermia
  - Heart block
  - Medication, e.g. β-blockers
  - CNS depression

Level of consciousness or new confusion Use the ACVPU score:

- Is the patient Alert and responding normally?
- Is the patient responding but Confused or disorientated? New-onset or worsening confusion/delirium warrants urgent clinical evaluation. If unsure whether confusion is 'new', treat as new until information about pre-morbid state becomes available
- Does the patient respond to Vocal stimuli?

- Does the patient respond to a **P**ainful stimulus (e.g. pinching the lower part of the nasal septum)?
- Is the patient **U**nconscious?

Temperature Both pyrexia and hypothermia are markers of acute illness severity, sepsis, and physiological disturbance.

Using the NEWS2 Add the score for the patient up.

- **Score 1–4—threat** Risk of deterioration. Consider whether acute referral is needed or whether can be safely monitored at home
- **Score 5 or 6 overall, or score of 3 in any specific parameter—refer** Unwell; needs urgent medical review in an acute care setting
- **Score ≥7—severe** Very unwell and needs emergency (blue light) referral to an acute care setting

❗ Actions must be appropriate to a patient's care plan, e.g. acute delirium in an elderly patient may be better managed at home.

⚠ Clinical concern about a patient should prompt action regardless of the NEWS2 score.

Physiological parameter	Score						
	3	2	1	0	1	2	3
Respiration rate (per minute)	≤8		9–11	12–20		21–24	≥25
SpO_2 Scale 1 (%)	≤91	92–93	94–95	≥96			
SpO_2 Scale 2 (%)	≤83	84–85	86–87	88–92 ≥93 on air	93–94 on oxygen	95–96 on oxygen	≥97 on oxygen
Air or oxygen?		Oxygen		Air			
Systolic blood pressure (mmHg)	≤90	91–100	101–110	111–219			≥220
Pulse (per minute)	≤40		41–50	51–90	91–110	111–130	≥131
Consciousness				Alert			CVPU
Temperature (°C)	≤35.0		35.1–36.0	36.1–38.0	38.1–39.0	≥39.1	

Figure 29.1 NEWS2 score

Reproduced from: Royal College of Physicians. *National Early Warning Score (NEWS) 2: Standardising the assessment of acute-illness severity in the NHS*. Updated report of a working party. London: RCP, 2017.

Further information

NHS Education for Scotland National Early Warning Score and Sepsis Screening Tool App. 🖱 www.knowledge.scot.nhs.uk/home/portals-and-topics/sepsis-app.aspx

Royal College of Physicians National Early Warning Score (NEWS) 2. 🖱 www.rcplondon.ac.uk/projects/outputs/national-early-warning-score-news-2

Managing a resuscitation attempt outside hospital

⚠ It is unacceptable for patients who sustain a cardiopulmonary arrest to await arrival of an ambulance before basic resuscitation or a defibrillator is available.

Resuscitation equipment See Box 29.2.

- Resuscitation equipment is used infrequently. It should be kept at strategic and accessible sites and not in a locked room/cupboard
- Staff must know where to find resuscitation equipment and be trained to use it to a level appropriate to the individual's expected role
- Emergency drugs should be kept in a box labelled 'for emergency use' which should be tamper-evident (e.g. sealed with tearable paper seals); if opened, the contents of the box should be checked before re-sealing
- Each practice should have a named individual with responsibility for checking the state of readiness of all resuscitation drugs and equipment on a regular basis, ideally daily. In common with drugs, disposable items like the adhesive electrodes have a finite shelf life and will require replacement from time to time if unused

Training Training and practice are necessary to acquire skills in resuscitation techniques. Resuscitation skills decline rapidly and updates are necessary every 6–12mo. Level of skill needed by different members of the primary healthcare team differs according to the individual's role:

- All those in direct contact with patients should be trained in basic life support and related resuscitation skills such as the recovery position
- Doctors, nurses, and other healthcare professionals should also be able to use an automatic external defibrillator (AED) effectively. Other personnel (e.g. receptionists) may also be trained to use an AED

❗ *In situ* simulation provides opportunities for the whole practice team to run through emergency scenarios safely within their own practice environment.

Performance management Records of all resuscitation attempts and electronic data stored by most AEDs during a resuscitation attempt should be kept for audit, training, and medicolegal reasons. Responsibility rests with the most senior member of the practice team involved. Process/outcome of all resuscitation attempts should be audited at practice and PCO level to allow deficiencies to be addressed and good practice to be shared.

Do not attempt to resuscitate (DNAR) decisions It is essential to identify individuals in whom cardiopulmonary arrest is a terminal event and where resuscitation is inappropriate. Overall responsibility for a DNAR decision rests with the clinician in charge of the patient's care:

- Seek opinions of other members of the medical and nursing team, the patient, and any relatives in reaching a DNAR decision
- Record the DNAR decision in the patient's notes, the reasons for that decision, and what the relatives have been told; provide a copy of the DNAR form for the patient/carers to keep in the home
- Ensure team members involved with the patient's care are aware of the decision. ❗ Notify the local ambulance service/OOH provider
- Review DNAR decisions regularly in the light of the patient's condition

Box 29.2 Suggested resuscitation equipment

- **Automated external defibrillator (AED) with electrodes and razor** Should be available wherever/whenever sick patients are seen. Preferably should have adult and paediatric settings. Regular maintenance is needed even if not used. After use, follow manufacturer's instructions to return the AED to a state of readiness with minimum delay
- **Pocket mask (adult) + oxygen port**
- **Oro-pharyngeal airway** For use by those appropriately trained. Keep a range of sizes (e.g. 0, 1, 2, 3, 4). Consider supraglottic airway device if trained to use
- **Oxygen, tubing, and mask with self-inflating reservoir bag** Ensure equipment is available in a range of sizes for both adults and children. Oxygen cylinders need regular maintenance—follow national safety standards
- **Suction** Portable, handheld suction devices are recommended
- **Drugs**
  - Adrenaline for cardiac arrest—adult dose is 1mg (10mL pre-filled syringes of 1 in 10,000 adrenaline are available)—ensure enough adrenaline to last until ambulance arrives (1mg every 4–5min)
  - Adrenaline for anaphylaxis—adult dose is 500 micrograms (0.5mL of 1 in 1000 adrenaline)
  - Chlorphenamine 10mg/mL
  - Hydrocortisone 100mg/mL ×2
  - Salbutamol 5mg vials for nebulisation ×5
  - Phenoxymethylpenicillin 1.2g (+ water for injection)
  - Glucogel®
  - IM glucagon 1mg/mL
  - Cefotaxime 1g
  - Diazepam (rectal) 10mg
- **Other:**
  - Algorithms, emergency drug doses, paediatric drug calculators
  - Protective equipment—gloves, aprons, eye protection
  - Pulse oximeter, stethoscope, BP monitor
  - Absorbent paper towel, 2% chlorhexidine/alcohol wipes
  - IV equipment—tourniquet; IV cannulae (variety of sizes); IV infusion set; sodium chloride 0.9% (2× 1000mL); glucose 10% (500mL)
  - Needles/syringes ± intraosseous device (if appropriate training)
  - Sharps box
  - Glucometer, lancets, and testing strips
  - Scissors
  - Tape

Further information

iResus Resuscitation algorithms. 🖱 https://www.resus.org.uk/apps/iresus/

Life-saver 🖱 www.life-saver.org.uk

Resuscitation Council (UK) (2013, updated 2017) Primary care—quality standards. 🖱 www.resus.org.uk/quality-standards/primary-care-quality-standards-for-cpr

Resuscitation Council (UK) (2013, updated 2015) Primary care—minimum equipment and drug lists for cardiopulmonary resuscitation. 🖱 www.resus.org.uk/quality-standards/primary-care-equipment-and-drug-lists

Resuscitation Council (UK) (2015) Prevention of cardiac arrest and decisions about CPR. 🖱 www.resus.org.uk/resuscitation-guidelines/prevention-of-cardiac-arrest-and-decisions-about-cpr

Adult basic life support

Paediatric basic life support ➔ p. 1044

Adult basic adult life support (ABLS) Is a holding operation—sustaining life until help arrives. ABLS should be started as soon as the arrest is detected—outcome is less good the longer the delay (Figure 29.2).

1. Danger Ensure safety of rescuer and patient
2. Response Check the patient for any response
   - Is the patient **Alert**? *Yes/No*
   - Is the patient **Confused** or disorientated? *Yes/No*
   - Does the patient respond to **Vocal** stimuli? *Yes/No*
   - Does the patient respond to a **Painful** stimulus (pinching the trapezius muscle between the shoulder and the neck)? *Yes/No*
   - Is the patient **Unconscious**? *Yes/No*

If the patient responds by answering or moving Do not move the patient unless in danger. Get help. Reassess regularly.

If the patient does not respond Shout for help; turn the patient onto his/her back.

3. Airway Open the airway—place one hand on the patient's forehead and tilt his/her head back. With fingertips under the point of the chin, lift the chin to open the airway.

⚠ Try to avoid head tilt if trauma to the neck is suspected.

4. Breathing With airway open, look, listen, and feel for breathing for no more than 10s—look for chest movement, listen at the patient's mouth for breath sounds, feel for air on your cheek.

If breathing normally Turn the patient into the recovery position (Figure 29.4, ➔ p. 1043), get help, and check for continued breathing.

❗ In the first few minutes after cardiac arrest, a patient may be barely breathing or taking infrequent, noisy, gasps. Do not confuse this with normal breathing. If any doubt whether breathing is normal, act as if it is *not* normal.

If not breathing Or only making occasional gasps/weak attempts at breathing, call for help. If you are the only rescuer, call for an emergency ambulance before starting CPR. If possible stay with the patient. If >1 rescuer is available, one should start chest compressions, while the other rescuer calls for an ambulance and fetches an AED (if available).

5. Circulation Start chest compressions if not breathing:
   - Kneel by the side of the patient and place the heel of one hand in the centre of the patient's chest. Place the heel of your other hand on top of the first hand and interlock the fingers of the two hands. Ensure pressure is not applied over the ribs, upper abdomen or lower end of the bony sternum
   - Position yourself vertically above the patient's chest and, with arms straight, press down on the sternum 5–6cm
   - After each compression, release all the pressure on the chest without losing contact between your hands and the sternum. Compression and release should take an equal amount of time
   - Repeat at a rate of ~100–120×/min

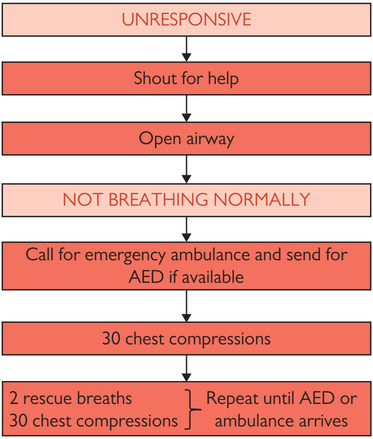

UNRESPONSIVE
Shout for help
Open airway
NOT BREATHING NORMALLY
Call for emergency ambulance and send for AED if available
30 chest compressions
2 rescue breaths
30 chest compressions
Repeat until AED or ambulance arrives

Figure 29.2 Adult basic life support (ABLS) algorithm

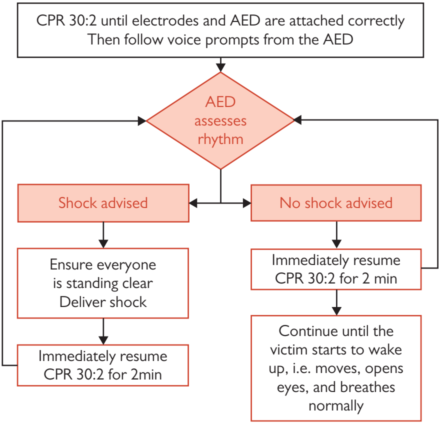

CPR 30:2 until electrodes and AED are attached correctly
Then follow voice prompts from the AED
AED assesses rhythm
Shock advised
No shock advised
Ensure everyone is standing clear
Deliver shock
Immediately resume CPR 30:2 for 2min
Immediately resume CPR 30:2 for 2 min
Continue until the victim starts to wake up, i.e. moves, opens eyes, and breathes normally

Figure 29.3 Automated external defibrillator algorithm

Source: data from Adult basic life support and automated external defibrillation, https://www.resus.org.uk/resuscitation-guidelines/adult-basic-life-support-and-automated-external-defibrillation/

⚠ Fatigue ↓ effectiveness of chest compressions after ~2min. If there is ≥1 trained CPR provider, change over every 2min to maintain effectiveness of CPR. Do not interrupt chest compressions while changing over. Ensure PPE is put on before attempting resuscitation of patients with high infection risk.

6. Combine chest compression with rescue breaths
    - After 30 compressions open the airway using head tilt and chin lift
    - Pinch the soft part of the patient's nose closed, using the index finger and thumb of your hand on the patient's forehead. Allow the patient's mouth to open, but maintain chin lift
    - Give a rescue breath—take a normal breath and place your lips around the patient's mouth (mouth-to-nose/mouth-to-tracheostomy are alternatives) making sure that there is a good seal. Blow steadily into the mouth for ~1s while watching the chest rise
    - Maintaining head tilt and chin lift; take your mouth away from the patient and watch for the chest to fall as air comes out then give a second rescue breath. Then return without delay to give a further 30 chest compressions
    - Continue chest compressions and rescue breaths in a ratio of 30:2

Using a pocket mask Ensure good seal around the patient's mouth/nose and give rescue breaths through the mask watching for chest rise.

Using a bag and mask Ensure good seal around the patient's nose/mouth and squeeze the bag firmly, watching the chest rise.

If rescue breaths do not make the chest rise

- Check the patient's mouth, and remove any visible obstruction
- Recheck that there is adequate head tilt and chin lift
- *Only attempt* 2 breaths each time before returning to chest compressions

⚠ Only stop CPR to reassess if the patient shows signs of regaining consciousness (e.g. coughs, opens eyes, moves purposefully) AND starts to breathe normally—otherwise resuscitation should not be interrupted.

Chest-compression-only CPR If unable/unwilling to give rescue breaths, give continuous chest compressions only at a rate of 100–120/min.

Use of automated external defibrillators (AEDs) in adults AEDs are programmed to deliver a single shock followed by a pause of 2min for the immediate resumption of CPR.

If a patient arrests Unless an AED is immediately available, start CPR according to ABLS guidelines.

As soon as the AED arrives See Figure 29.3, ➔ p. 1041.

- Switch on the AED and attach the electrode pads. If >1 rescuer is present, continue CPR while this is done (some AEDs automatically switch on when the AED lid is opened)
    - Place one AED pad to the right of the sternum below the clavicle
    - Place the other pad in the left mid-axillary line with its long axis vertical
- Follow the voice/visual prompts. Ensure nobody touches the patient while the AED is analysing the rhythm

If a shock is indicated Ensure that nobody touches the patient. Push the shock button as directed (fully automatic AEDs deliver the shock automatically). Immediately resume CPR and continue to follow the prompts.

If no shock is indicated Immediately resume CPR and continue to follow the voice prompts from the AED.

Use of AEDs in children ➔ p. 1046

Adrenaline (epinephrine) and oxygen

- If able to gain IV access and adrenaline is available, give 1mg adrenaline immediately and every 3–5min thereafter or after alternate AED cycles
- If oxygen is available, give 100% high-flow oxygen. Ensure mask and oxygen tubing is >1m from the patient's chest when attempting defibrillation

Duration of resuscitation Continue resuscitation until:

- Further qualified help arrives from the emergency medical services
- The victim starts breathing normally, *and/or*
- You become exhausted

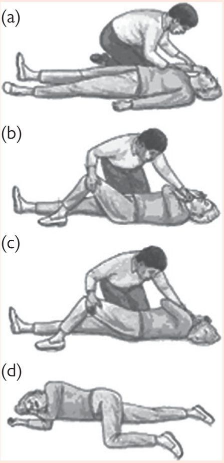
Management after successful treatment of cardiac arrest

- Turn into the recovery position (Figure 29.4)
- Give oxygen aiming to keep peripheral oxygen saturation at 94–98%
- Transfer to hospital as soon as possible by emergency ambulance

Figure 29.4 The recovery position. When a patient is unconscious but has circulation and is breathing, it is important to maintain a good airway; ensure the tongue does not cause obstruction; and minimize the risk of inhalation of gastric contents.

Putting a patient into the recovery position

- Remove the patient's glasses
- Kneel beside the patient and make sure both legs are straight (a)
- Place the arm nearest to you out at right angles to the body, elbow bent with the hand palm uppermost (a)
- Bring the far arm across the chest, and hold the back of the hand against the patient's cheek nearest to you (b)
- With your other hand, grasp the far leg just above the knee and pull it up, keeping the foot on the ground (b)
- Keeping the patient's hand pressed against their cheek, pull on the leg to roll the patient towards you onto their side (c)
- Adjust the upper leg so that both the hip and knee are bent at right angles (d)
- Tilt the head back to ensure the airway remains open (d); adjust hand under the cheek, if needed, to keep the head tilted
- Check breathing regularly



⚠ Monitor the peripheral circulation of the lower arm. If the patient is in the recovery position for >30min, turn the patient onto the opposite side.

Further information

Resuscitation Council (UK) (2015) Prehospital resuscitation. 🖱 www.resus.org.uk/resuscitation-guidelines/prehospital-resuscitation/

Resuscitation Council (UK) (2015) Adult basic life support and automated external defibrillation. 🖱 www.resus.org.uk/resuscitation-guidelines/adult-basic-life-support-and-automated-external-defibrillation/

Paediatric basic life support

Adult basic life support ➔ p. 1040

Resuscitation of the newborn ➔ p. 1086

Paediatric basic life support (PBLS) Is a holding operation—sustaining life until help arrives. PBLS should be started as soon as the arrest is detected—outcome is less good the longer the delay (Figure 29.5).

1. Danger Ensure safety of rescuer and child
2. Response Check the child for any response
    - Is the child **Alert**? *Yes/No*
    - Is the child **Confused** or disorientated? *Yes/No*
    - Does the child respond to **Vocal** stimuli? *Yes/No*
    - Does the child respond to a **Painful** stimulus (pinching the trapezius muscle between the neck/shoulder)? *Yes/No*
    - Is the child **Unconscious**? *Yes/No*

If the child responds by answering or moving Do not move the child unless in danger. Get help. Reassess regularly.

If the child does not respond Shout for help; turn the child onto his/her back.

3. Airway Open the airway:
    - Gently tilt the head back—with your hand on the child's forehead
    - Lift the chin—with your fingertips under the point of the child's chin
    - Do not push on the soft tissues under the chin
    - *If unsuccessful* Try jaw thrust—place the first 2 fingers of each hand behind each side of the child's jaw bone, and push the jaw forward
4. Breathing Look, listen, and feel for breathing (maximum 10s)

⚠ Avoid head tilt as much as possible if trauma to the neck is suspected.

If breathing normally Turn the child carefully into the recovery position (➔ p. 1043) if unconscious, and check for continued breathing.

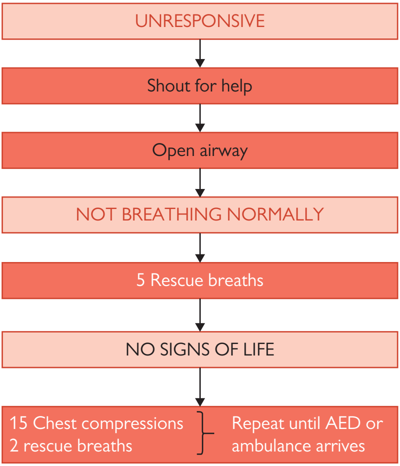
If not breathing Or not breathing normally:

- Remove any obvious airway obstruction
- Give 5 initial rescue breaths—note any gag or cough response

Technique for rescue breaths

- Ensure head tilt (neutral position for children <1y) and chin lift
- If age ≥1y, pinch the soft part of the child's nose closed with the index finger and thumb of the hand which is on their forehead. Open the child's mouth a little, but maintain the chin upwards
- Take a breath and place your lips around the child's mouth (mouth and nose if <1y), ensuring you have a good seal. Blow steadily into the child's airway over ~1s, watching for the chest to rise
- Maintaining head tilt and chin lift, take your mouth away and watch for the chest to fall as air comes out
- Take another breath and repeat this sequence 5 times

❗ Use paediatric bag-mask device if available. If you have difficulty achieving an effective breath, consider airway obstruction—➔ p. 1050.


UNRESPONSIVE
Shout for help
Open airway
NOT BREATHING NORMALLY
5 Rescue breaths
NO SIGNS OF LIFE
15 Chest compressions
2 rescue breaths
Repeat until AED or ambulance arrives

If lone rescuer, after 1 min call for ambulance and then continue CPR

Figure 29.5 Paediatric basic life support algorithm

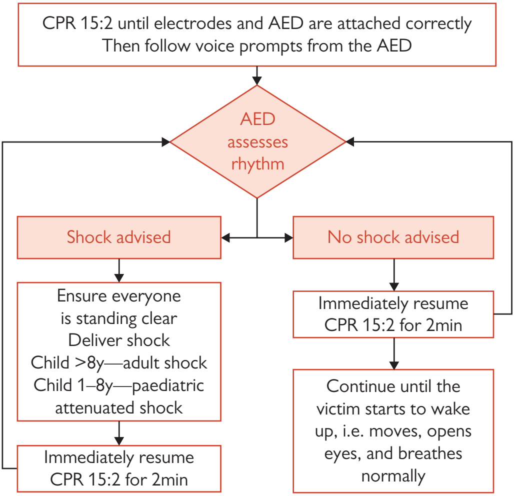

CPR 15:2 until electrodes and AED are attached correctly
Then follow voice prompts from the AED
AED assesses rhythm
Shock advised
No shock advised
Ensure everyone is standing clear
Deliver shock
Child >8y—adult shock
Child 1–8y—paediatric attenuated shock
Immediately resume CPR 15:2 for 2min
Immediately resume CPR 15:2 for 2min
Continue until the victim starts to wake up, i.e. moves, opens eyes, and breathes normally

Figure 29.6 Paediatric AED algorithm

Source: data from Paediatric advanced life support, https://www.resus.org.uk/resuscitation-guidelines/paediatric-advanced-life-support/

5. **C**irculation (signs of life) Check (maximum 10s) for:
    - Any movement, coughing, or normal breathing (not agonal gasps)
    - Pulse—child ≥1y carotid/femoral pulse; child <1y brachial pulse

If circulation is present Continue rescue breathing until the child starts breathing effectively on their own. Turn the child into the recovery position (➲ p. 1043) if unconscious, and reassess frequently.

If circulation is absent Or slow pulse (<60bpm) with poor perfusion or you are not sure:

- Give 15 chest compressions. Then give 2 rescue breaths, followed by 15 further chest compressions
- Continue the cycle of 2 breaths followed by 15 chest compressions

❗ Lone rescuers may use a ratio of 30 compressions to 2 rescue breaths.

Technique for chest compressions Compress the sternum 1 finger's breadth above the xiphisternum by at least a third of the depth of the chest. Release the pressure then repeat at a rate of ~100–120 compressions/min.

- Children <1y with a lone rescuer Use the tips of 2 fingers
- Children <1y with ≥2 rescuers Place both thumbs flat on the lower third of the sternum with tips pointing towards the child's head and encircle the lower part of the child's ribcage with the tips of the fingers supporting the infant's back. Press down with both thumbs
- Children >1y Place the heel of 1 hand over the lower half of the sternum. Lift the fingers. Position yourself vertically above the chest with arm straight, and push downwards. For larger children use both hands with fingers interlocked to achieve satisfactory compressions

⚠ Stop to recheck for signs of a circulation only if the child moves or takes a spontaneous breath—otherwise continue uninterrupted.

When to call for emergency ambulance support It is vital for rescuers to get assistance as quickly as possible when a child collapses.

When >1 rescuer is available One rescuer should start resuscitation while another rescuer calls for emergency ambulance support and fetches an AED if available.

Lone rescuer Perform resuscitation for 1min before calling for emergency ambulance support. The only exception to this is a witnessed sudden collapse—as in this case cardiac arrest is likely to be due to arrhythmia and the child may need defibrillation, so seek help immediately

Use of automated external defibrillators (AEDs) in children

- Children >8y Use the standard adult AED
- Children <8y Paediatric pads or a paediatric mode should be used if available—if not, use the adult AED as it is

❗ AEDs are not designed for use in infants <1y. Shockable rhythm is rare in children <1y but, if present, benefits of AED use (preferably using attenuated paediatric setting) outweigh risks.

If a child arrests Start CPR in accordance with PBLS guidelines.

As soon as the AED arrives Follow Figure 29.6, ➔ p. 1045.

- Switch on the AED and attach the electrode pads. If >1 rescuer is present, continue CPR while this is done. (Some AEDs automatically switch on when the AED lid is opened)
- Place one AED pad to the right of the sternum below the clavicle
- Place the other pad in the mid-axillary line with its long axis vertical
- Follow the voice/visual prompts. Ensure nobody touches the child while the AED is analysing the rhythm

If a shock is indicated Ensure nobody touches the child. Deliver the shock as directed by the voice prompt.

Adrenaline (epinephrine) and oxygen

- IV access is difficult to obtain in children; consider intraosseous access if equipment is available and suitably trained. If adrenaline is available, give 10 micrograms/kg adrenaline (0.1mL/kg of 1:10,000 solution) every 3–5min or after alternate AED cycles. Use a recent weight from the parent-held record—otherwise, for children aged >1y, weight (in kg) ≈2× (age + 4)
- If oxygen is available, give 100% high-flow oxygen. Ensure mask and oxygen tubing is >1m from the child's chest if attempting defibrillation

Duration of resuscitation Continue resuscitation until:

- Child shows signs of life (spontaneous respiration, pulse, movement)
- Further qualified help arrives from emergency medical services
- You become exhausted

Cervical spine injury

- If spinal cord injury is suspected (e.g. if the child has sustained a fall, been struck on the head or neck, or has been rescued after diving into shallow water) take particular care during handling and resuscitation to maintain alignment of the head, neck, and chest in the neutral position
- A spinal board and/or cervical collar should be used if available

Management after successful treatment of cardiac arrest

- Turn into the recovery position
- Give oxygen, aiming to keep oxygen saturation at 94–98%
- Keep the child warm
- Transfer to hospital as soon as possible by emergency ambulance

The recovery position for an unconscious child See Figure 29.4, ➔ p. 1043. The child should be in as near a true lateral position as possible with his/her mouth dependent to allow free drainage of fluid. The position should be stable. In an infant this may require the support of a small pillow or rolled-up blanket placed behind the infant's back to maintain the position.

Further information

Resuscitation Council (UK) (2015) Prehospital resuscitation. 🖱 www.resus.org.uk/resuscitation-guidelines/prehospital-resuscitation/

Resuscitation Council (UK) (2015) Paediatric basic life support. 🖱 www.resus.org.uk/resuscitation-guidelines/paediatric-basic-life-support/

The choking adult

⚠ If blockage of the airway is only partial, the patient will usually be able to dislodge the foreign body by coughing. If obstruction is complete, urgent intervention is required to prevent asphyxia (Figure 29.7).

Is foreign body airways obstruction (FBAO) likely?

- Sudden onset of respiratory distress while eating?
- Is the patient clutching his/her neck?

Is the patient coughing effectively?

Signs of an effective cough include

- Patient responds 'Yes' to the question '*Are you choking?*'
- Fully responsive—able to speak, cough, and breathe

►► Encourage the patient to cough, and monitor.

Signs of an ineffective cough include

- In response to the question '*Are you choking?*' the patient either responds by nodding or is unable to respond
- Breathing sounds wheezy
- Unable to breathe
- Attempts at coughing are silent
- Unconscious

►► Call for an emergency ambulance and assess conscious level.

If the patient is conscious but has absent/ineffective cough

- Give up to 5 back blows as needed
- If back blows do not relieve the obstruction, give up to 5 abdominal thrusts

Following back blows or abdominal thrusts Reassess:

- If the object has not been expelled and the victim is still conscious Continue the sequence of back blows and abdominal thrusts
- If the object is expelled successfully Assess clinical condition (including abdominal examination if abdominal thrusts used). If there is any suspicion part of the object is still in the respiratory tract or any intra-abdominal injuries as a result of abdominal thrusts, refer to A&E for assessment

If the victim becomes unconscious

- Support the patient carefully to the ground
- Immediately call for ambulance support
- Begin CPR (➔ p. 1040), with 30 chest compressions at a rate of 100–120/min—even if carotid pulse is present

Foreign body in the throat Occurs after eating—fish bone or food bolus are most common. Can cause severe discomfort, distress, and inability to swallow saliva.

Management Refer immediately to A&E or ENT for investigation (lateral neck X-ray ± laryngoscopy). Most fish bones have passed and discomfort comes from mucosal trauma. Food boluses often pass (especially if the patient is given a smooth muscle relaxant) but occasionally need removal under GA.

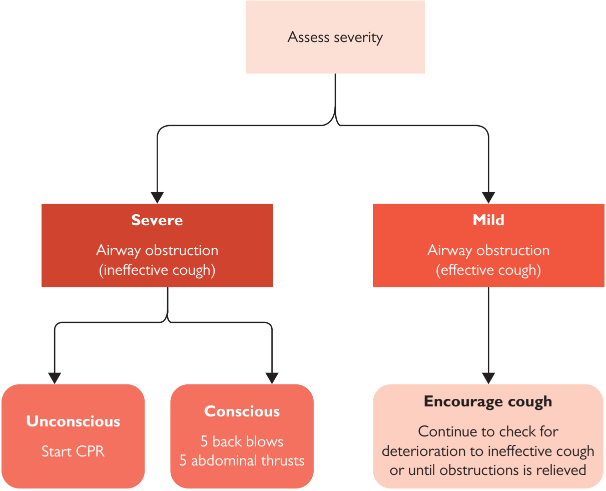

Assess severity
Severe
Airway obstruction (ineffective cough)
Mild
Airway obstruction (effective cough)
Unconscious
Start CPR
Conscious
5 back blows
5 abdominal thrusts
Encourage cough
Continue to check for deterioration to ineffective cough or until obstructions is relieved

Figure 29.7 Algorithm for management of choking in adults

Reproduced with the kind permission of the Resuscitation Council (UK).

Back blows for adults

- Stand to the side and slightly behind the patient
- Support the chest with 1 hand and lean the patient well forwards so that when the obstructing object is dislodged it comes out of the mouth
- Give up to 5 sharp blows between the shoulder blades with the heel of the other hand

Abdominal thrusts for adults

- Stand behind the patient and put both arms around the upper part of the abdomen
- Lean the patient forwards
- Clench your fist and place it between the umbilicus and bottom end of the sternum
- Grasp this hand with your other hand and pull sharply in- and upwards
- Repeat up to 5 times as needed

Further information

Resuscitation Council (UK) (2015) Adult basic life support and automated external defibrillation. 🖱 www.resus.org.uk/resuscitation-guidelines/adult-basic-life-support-and-automated-external-defibrillation/

The choking child

⚠ If breathing spontaneously, encourage the child's own efforts to clear the obstruction. ONLY intervene if ineffective (Figure 29.8).

Is foreign body airways obstruction (FBAO) likely? Look for:

- Sudden onset of respiratory distress in a previously well child—often witnessed by the child's carer
- Respiratory distress associated with coughing, gagging, or stridor
- Recent history of playing with or eating small objects

Is the child coughing effectively?

Signs of an effective cough include

- Fully responsive—crying or verbal response to questions
- Loud cough and able to take a breath before coughing

►► Encourage the child to cough and monitor.

Signs of an ineffective cough include

- Unable to vocalize
- Quiet or silent cough
- Unable to breathe ± cyanosis
- Decreasing level of consciousness

►► Call for an emergency ambulance and assess conscious level.

If the child is conscious but has absent/ineffective cough

- Give up to 5 back blows as needed. If back blows do not relieve the obstruction, give up to 5 chest thrusts (infants <1y) or up to 5 abdominal thrusts (children ≥1y) as needed. Then reassess
- **If the object has not been expelled and the child is still conscious** Continue the sequence of back blows and chest (for infant) or abdominal (for child aged >1y) thrusts. ❗ Do not leave the child
- **If the object is expelled successfully** Assess clinical condition (including abdominal examination if abdominal thrusts used). If there is any suspicion part of the object is still in the respiratory tract or there are any intra-abdominal injuries as a result of abdominal thrusts, refer to A&E

If the child is unconscious ❗ Do not leave the child.

- Place on a firm, flat surface—call out/send for help if not arrived
- Open the mouth and look for any obvious object. If one is seen, make an attempt to remove it with a single finger sweep
- Open the airway and attempt 5 rescue breaths. Assess effectiveness of each breath—if a breath does not make the chest rise, reposition the head before making the next attempt
- If there is no response to rescue breaths, proceed immediately to chest compression—regardless of whether the breaths were successful. Follow the PBLS sequence (➔ p. 1044) for 1min before summoning help (if not already there)

If it appears the obstruction has been relieved

- Open and check the airway. Deliver rescue breaths if not breathing
- If the child regains consciousness and is breathing effectively, place in a safe side-lying (recovery) position, and monitor breathing and conscious level while awaiting the arrival of the emergency services

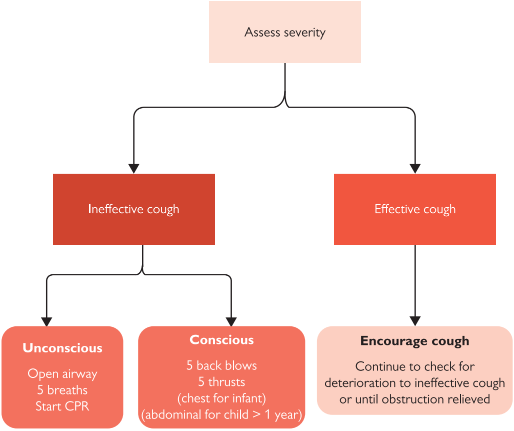

Assess severity
Ineffective cough
Effective cough
Unconscious
Open airway
5 breaths
Start CPR
Conscious
5 back blows
5 thrusts
(chest for infant)
(abdominal for child > 1 year)
Encourage cough
Continue to check for deterioration to ineffective cough or until obstruction relieved

Figure 29.8 Algorithm for management of paediatric foreign body airways obstruction
Reproduced with the kind permission of the Resuscitation Council (UK).

Back blows for small children/infants

- Place the child in a head-downwards, prone position (e.g. across your lap). Support the head if needed by holding the jaw
- Deliver a smart blow with the heel of one hand to the middle of the back between the shoulder blades. Repeat up to 5 times as needed

Back blows for older children

- Support the child in a forward-leaning position
- Deliver a smart blow with the heel of one hand to the middle of the back between the shoulder blades from behind. Repeat up to 5 times as needed

Chest thrusts for infants <1y

- Turn the child into a supine position with head down (e.g. by holding the child's occiput and laying the child along your arm, supported on your thigh)
- Deliver 5 sharp chest thrusts (like chest compressions but slower rate ~20/min) to a point 1 finger's breadth above the xiphisternum

Abdominal thrusts for children ≥1y

- Stand behind the child (kneel if small child). Place your arms under the child's arms and encircle his torso
- Clench your fist and place it between the umbilicus and xiphisternum
- Grasp your clenched hand with your other hand and pull sharply inwards and upwards. Repeat up to 5 times as needed

❗ Ensure that pressure is not applied to the xiphoid process or the lower rib cage as this may cause abdominal trauma.

Further information

Resuscitation Council (UK) (2015) Paediatric basic life support. 🔗 www.resus.org.uk/resuscitation-guidelines/paediatric-basic-life-support/

The unconscious patient

⚠ If you receive a call for assistance, advise the attendant to:

- Call for an emergency ambulance *and*
- Turn the patient onto his/her side (unless history of possible spinal injury when should await emergency services before moving)

Assessment of an unconscious patient

- Establish that the patient is unconscious—shake the patient and ask '*Are you alright?*'
- Ensure safety e.g. stop traffic, put on gloves
- Call for help from colleagues/bystanders and emergency services (if not already called) as soon as possible
- Do a complete initial assessment and re-assess regularly
- Use an ABCDE approach—Figure 29.9
- Obtain additional information if possible, e.g. history from attendant, check medical notes for past medical history, clues from the scene (e.g. suicide note, used needles/drug injecting equipment)
- Record observations each time assessed; calculate NEWS score (➔ p. 1036); communicate effectively, e.g. using SBAR (➔ p. 1035)
- Keep the patient warm

If equipment is available

- Insert an airway
- Gain IV access
- Attach BP monitor, pulse oximeter ± ECG monitor/AED
- Give oxygen (15L/min) to maintain peripheral oxygen saturations at 94–98% (unless known hypercapnic respiratory failure, when aim for 88–92%)

Transient loss of consciousness ➔ p. 524

Syncope and risk of sudden cardiac death Refer to cardiology for assessment in <24h if[N]:

- Any abnormalities on ECG
- New/unexplained breathlessness
- Heart failure (history/clinical signs)
- Heart murmur
- Transient loss of consciousness during exertion
- Family history of sudden cardiac death <40y and/or inherited heart condition

Consider referral if atypical syncope (e.g. syncope lying down) or ≥65y and transient loss of consciousness without prodromal symptoms.

The fitting patient ➔ p. 1080

Table 29.1 The Glasgow coma scale

Eye opening	Spontaneous	4	To pain	2
	To voice	3	None	1
Best verbal response	Oriented	5	Incomprehensible	2
	Confused	4	None	1
	Inappropriate words	3		
Best motor response	Obeys commands	6	Flexion	3
	Localizes pain	5	Extension	2
	Withdraws from pain	4	None	1

Total score = Eye opening + Best verbal response + Best motor response scores.

Reprinted from *The Lancet*, **13**, Teasdale G, et al., Assessment of coma and impaired consciousness, 81–4, Copyright (1974), with permission from Elsevier.

A—AIRWAY	**Does the patient have a patent airway?** • Look for signs of airway obstruction e.g. paradoxical ('see-saw' chest and abdominal movements; lack of air movement from mouth/nose; use of accessory muscles; stridor (partial airway obstruction) • If airway is obstructed, try opening the airway using head tilt and jaw thrust—➲ p. 1040; give high flow oxygen if available • Consider choking—➲ p. 1048 (adult) and ➲ p. 1050 (child) • If unconscious but has circulation and breathing, consider turning into the recovery position—Figure 29.4, ➲ p. 1043
B—BREATHING	**Is the patient breathing?** • If not, start ABLS (➲ p. 1040) or PBLS (➲ p. 1044) • If breathing, assess and record respiratory rate and peripheral oxygen saturation. Normal respiratory rate for an adult is 12–20 breaths/min. Respiratory rate ≥25 breaths/min is a marker of severe illness and warning sign for rapid deterioration • Look for signs of respiratory distress—use of accessory muscles, abdominal breathing, sweating, cyanosis • Examine the chest looking for tracheal deviation, ↓ air entry, added sounds, signs of pneumothorax • Consider giving high flow oxygen if available to maintain peripheral oxygen saturations between 94–98% (88–92% if hypercapnic respiratory failure)
C—CIRCULATION	**Is the patient shocked?** ➲ p. 1060 • Check and record BP, pulse rate, and capillary refill time (>2sec is abnormal) • Look for evidence of sepsis, anaphylaxis, haemorrhage, and heart disease (rub, murmur, arrhythmia)
D—DISABILITY	• **Level of consciousness**—check and record ACVPU (➲ p. 1036) ± Glasgow coma scale (Table 29.1) • **Capillary blood glucose**—if <4mmol/L consider hypoglycaemia; if >7mmol/L consider hyperglycaemic state (➲ p. 1082)
E—EXPOSURE	**Look for clues for underlying cause of coma**, e.g. • MedicAlert® bracelet/jewellry • Signs of injury e.g. head injury (➲ p. 1094), haemorrhage • Smell of alcohol • Pin-point pupils/needle marks ?opioid overdose—➲ p. 1098 • Fever ± rash ? sepsis—➲ p. 1056 • Urticaria/angio-oedema ? anaphylaxis—➲ p. 1056 • Hypothermia

Figure 29.9 The ABCDE approach[G]

❗ Aims to keep the patient alive, and achieve clinical improvement. Treat life-threatening problems before moving to the next part of the assessment. Assess effects of treatment.

Further information

NICE (2010, updated 2014) Transient loss of consciousness ('blackouts') in over 16s. 🖱 www.nice.org.uk/guidance/cg109

Resuscitation Council (UK) The ABCDE approach. 🖱 www.resus.org.uk/resuscitation-guidelines/abcde-approach/

Anaphylaxis

Severe systemic allergic reaction that is life-threatening. *Common causes*:

- Foods Nuts, milk, fruit, fish and shellfish, eggs, pulses (beans, peas)
- Drugs Antibiotics, aspirin and other NSAIDs, opioids
- Insect stings Wasp or bee
- Latex

Features Often history of anaphylaxis/severe allergic reaction. Anaphylaxis is likely if sudden onset/rapid progression of symptoms (minutes) and life-threatening:

- Airway problems—difficulty breathing/swallowing; feeling that throat is closing; hoarseness; stridor; *and/or*
- Breathing problems—↑ respiratory rate; wheeze; shortness of breath; peripheral oxygen saturation <92%; cyanosis (late sign); confusion due to anoxia; respiratory arrest; *and/or*
- Circulation problems—shock (pallor, clammy, tachycardia—bradycardia is a late feature); ↓ BP; faintness/dizziness; collapse; agitation/confusion; loss of consciousness. May cause myocardial ischaemia and ECG changes even if normal coronary arteries

Skin and/or mucosal changes May be subtle/absent in 1 in 5. *Include*:

- Flushing
- Erythema
- Urticaria and/or angio-oedema
- Rhinitis and/or conjunctivitis

❗ Skin/mucosal changes alone are not a sign of an anaphylaxis.

Other symptoms Abdominal symptoms, e.g. abdominal pain, vomiting or incontinence; anxiety ± sense of impending doom.

Differential diagnosis

- Life-threatening Severe asthma; septic shock
- Non-life-threatening Simple faint; hyperventilation/panic attack; breath-holding attacks in small children; lone urticaria/angio-oedema

If telephone contact Request an emergency ambulance immediately. Ask if the patient has had a similar event before. If so, does he/she have an adrenaline auto-injector device? If yes, advise immediate use.

If with the patient Follow the algorithm in Figure 29.10.

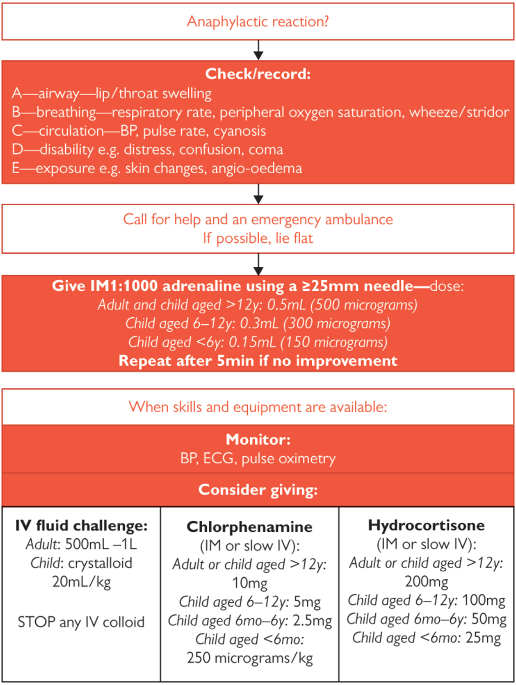
- Patients with airway/breathing problems may prefer to sit up
- If ↓ BP, lie flat (on left side if pregnant) with legs elevated
- If unconscious and breathing, place in the recovery position (Figure 29.4, ➔ p. 1043)

Follow-up

- Admit to A&E, even if seemingly recovered; symptoms may recur
- Warn of the possibility of future recurrence—advise to wear a tag (e.g. Medic-Alert bracelet 🖱 www.medicalert.org.uk) to alert others
- Refer all patients to a specialist allergy clinic after first anaphylactic attack
- Supply an adrenaline auto-injector device (e.g. EpiPen®) to administer IM adrenaline (epinephrine) immediately should symptoms recur

Prescribing adrenaline auto-injectors 2 auto-injectors should be prescribed and carried by the patient at all times. Ensure patients/parents/carers have been trained to use the auto-injector prescribed—trainer devices to practise with are available from manufacturers. *Advise to*:

- Use at the first signs of severe allergic reaction, and to lie flat, elevating legs (unless breathing difficulty when should sit up)
- Seek help from others immediately after using the auto-injector and call for an emergency ambulance stating '*anaphylaxis*'—even if symptoms are improving; ensure someone is with the patient until the ambulance arrives
- If no improvement after 5–15min, use the second auto-injector
- Check the expiry date of auto-injectors and obtain replacements before they expire; expired injectors are less effective



Anaphylactic reaction?

Check/record:
A—airway—lip/throat swelling
B—breathing—respiratory rate, peripheral oxygen saturation, wheeze/stridor
C—circulation—BP, pulse rate, cyanosis
D—disability e.g. distress, confusion, coma
E—exposure e.g. skin changes, angio-oedema

Call for help and an emergency ambulance
If possible, lie flat

Give IM1:1000 adrenaline using a ≥25mm needle—dose:
Adult and child aged >12y: 0.5mL (500 micrograms)
Child aged 6–12y: 0.3mL (300 micrograms)
Child aged <6y: 0.15mL (150 micrograms)
Repeat after 5min if no improvement

When skills and equipment are available:

Monitor:
BP, ECG, pulse oximetry

Consider giving:

IV fluid challenge:	**Chlorphenamine** (IM or slow IV):	**Hydrocortisone** (IM or slow IV):
Adult: 500mL –1L	*Adult or child aged >12y*: 10mg	*Adult or child aged >12y*: 200mg
Child: crystalloid 20mL/kg	*Child aged 6–12y*: 5mg	*Child aged 6–12y*: 100mg
STOP any IV colloid	*Child aged 6mo–6y*: 2.5mg	*Child aged 6mo–6y*: 50mg
	Child aged <6mo: 250 micrograms/kg	*Child aged <6mo*: 25mg

Figure 29.10 Immediate response to a suspected anaphylactic reaction[G]

Source: data from Emergency treatment of anaphylactic reactions: Guidelines for healthcare providers, https://www.resus.org.uk/anaphylaxis/emergency-treatment-of-anaphylactic-reactions/

Further information

MHRA (2017) Adrenaline auto-injectors: updated advice after European review. 🖱 www.gov.uk/drug-safety-update/adrenaline-auto-injectors-updated-advice-after-european-review
Resuscitation Council (UK) (2012) Anaphylaxis. 🖱 www.resus.org.uk/anaphylaxis/emergency-treatment-of-anaphylactic-reactions

Information and support for patients

Allergy UK ☎ 01322 619898 🖱 www.allergyuk.org
Anaphylaxis Campaign ☎ 01252 542029 🖱 www.anaphylaxis.org.uk

Sepsis, meningitis, and encephalitis

Sepsis Defined as life-threatening organ dysfunction caused by a dysregulated host response to an infection. It can be triggered by any infection.

Initial assessment of patients with possible infection ➲ p. 622. Think '*Could this be sepsis?*'—particularly if the person looks ill or the family/carer is concerned.

Risk stratification for suspected sepsis

- Adults and children ≥12y Figure 29.11
- Children age 5–12y Figure 29.12, ➲ p. 1058
- Children age <5y Figure 29.13, ➲ p. 1059

Management

- If any red flag (high-risk) features (Figures 29.11, 29.12, and 29.13), admit via blue light ambulance to hospital. Give O_2 to maintain peripheral saturations >94% while awaiting transfer. Provide clear handover to paramedics (e.g. SBAR—➲ p. 1035; NEWS2 score—➲ p. 1036)
- If any amber flag features (Figures 29.11, 29.12, and 29.13), assess whether can be safely managed in the community, e.g. consider admission if lives alone. If managed in the community, treat underlying infection as appropriate. Provide clear safety netting advice about when/how to seek further advice if there is any deterioration or concern. Consider arranging early follow-up. ❗ If <17y and any immune suppression, admit

Neutropenic sepsis ➲ p. 626

Meningitis and encephalitis[N] Present in similar fashion. Usually rapid onset (<48h). Typical symptoms may be preceded by a prodrome of fever, vomiting, malaise, poor feeding, and lethargy which is often indistinguishable from a viral infection. Particularly significant early signs include:

- Severe leg pain—so bad that the child cannot stand/walk
- Cold hands or feet when the child is running a fever
- Pale skin ± blueness around the lips

Typical symptoms/signs

- Meningism Headache, stiff neck (cannot put chin on chest), and photophobia. Kernig's sign may be +ve (with hips fully flexed, resists passive knee extension). May also be seen with subarachnoid haemorrhage
- ↑ intracranial pressure Bulging fontanelle (babies); drowsiness/↓ level of consciousness; vomiting; irritability; ↓ pulse rate; ↑ BP; fitting; neurological signs, e.g. abnormal tone/posturing
- Septicaemia/septic shock Figures 29.11, 29.12, and 29.13. A petechial rash suggests meningococcal septicaemia

❗ Small children/immunocompromised patients may present atypically.

⚠ *Action* Call an emergency ambulance; transfer to hospital as soon as possible. If shocked, lie the patient flat and raise legs above waist height. If available, give 100% O_2 (15L/min). Gain IV access if possible.

If symptoms/signs of meningococcal disease Give IV/IM antibiotics immediately while awaiting transport—see Box 29.3. *Do not delay* hospital transfer to give antibiotics. For meningitis without signs of meningococcal disease, only give antibiotics if urgent hospital transfer is not possible.

Category	High risk criteria	Moderate to high risk criteria	Low risk criteria
History	Objective evidence of new altered mental state	History from patient, friend, or relative of new onset of altered behaviour or mental state History of acute deterioration of functional ability Impaired immune system (illness or drugs including oral steroids) Trauma, surgery, or invasive procedures in the last 6wk	Normal behaviour
Respiratory	↑ respiratory rate: ≥25 breaths/min New need for O_2 to maintain saturation >92% (or >88% if COPD)	↑ respiratory rate: 21–24 breaths/min	No high risk or moderate to high risk criteria met
BP	Systolic BP: ≤90mmHg or >40mmHg below normal	Systolic BP: 91–100mmHg	No high risk or moderate to high risk criteria met
Circulation/ hydration	↑ heart rate: >130 bpm Not passed urine in ≥18h (if catheterized, passed <0.5 ml/kg/h of urine)	↑ heart rate: 91–130 bpm (pregnant ♀: 100–130 bpm) or new onset arrhythmia Not passed urine in 12–18h (if catheterized, passed 0.5–1 ml/kg/h of urine)	No high risk or moderate to high risk criteria met
Temperature		Tympanic temperature <36°C	
Skin	Mottled or ashen appearance Cyanosis of skin, lips, or tongue Non-blanching skin rash	Signs of potential infection, including redness, swelling, or discharge at surgical site or breakdown of wound	No non-blanching rash

Figure 29.11 Risk stratification tool for adults and children ≥12y with suspected sepsis[N]

Reproduced with permission from NICE (2016, updated 2017) Sepsis: recognition, diagnosis and early management. www.nice.org.uk/guidance/ng51

Box 29.3 Antibiotics for suspected meningococcal sepsis

- Give benzylpenicillin. *Dose*: adult or child ≥10y—1.2g; child 1–9y—600mg; infant <1y—300mg
- Withhold penicillin ONLY if clear history of anaphylaxis in response to a previous dose. Rash following penicillin is not a contraindication
- Cefotaxime is an alternative: adult or child >12y—1g; child <12y—50mg/kg

Category	High-risk criteria	Moderate- to high-risk criteria	Low-risk criteria
Behaviour	Objective evidence of altered behaviour or mental state Appears ill to a healthcare professional Does not wake or, if roused, does not stay awake	Not behaving normally ↓ activity Parent/carer concern that the child is behaving differently from usual	Normal behaviour
Respiratory	O_2 saturation of <90% in air or ↑ O_2 requirement from baseline ↑ respiratory rate: 5y ≥29 breaths/min 6–7y ≥27 breaths/min 8–11y ≥25 breaths/min	O_2 saturation of <92% in air or ↑ O_2 requirement from baseline ↑ respiratory rate: 5y 24–28 breaths/min 6–7y 24–26 breaths/min 8–11y 22–24 breaths/min	No high-risk or moderate- to high-risk criteria met
Circulation/ hydration	Heart rate <60bpm ↑ heart rate: 5y ≥130bpm 6–7y ≥120bpm 8–11y ≥115bpm	Capillary refill time ≥3s ↑ heart rate: 5y 120–129bpm 6–7y 110–119bpm 8–11y 105–114bpm	No high-risk or moderate- to high-risk criteria met
Temperature		Tympanic temperature <36°C	No high-risk or moderate- to high-risk criteria met
Skin and other	Mottled or ashen appearance Cyanosis of skin, lips, or tongue Non-blanching skin rash	Leg pain Cold hands/feet	

Figure 29.12 Risk stratification tool for children age 5–12y with suspected sepsis[N]

Reproduced with permission from NICE (2016, updated 2017) Sepsis: recognition, diagnosis and early management. www.nice.org.uk/guidance/ng51

Contact tracing/prophylaxis for meningococcal disease Meningitis and acute encephalitis are notifiable diseases. Contact tracing is undertaken by the local public health department. For a single case, only close contacts ('kissing contacts'), e.g. people living in the same household, require prophylactic antibiotics and vaccination. *Prophylactic antibiotics*:

- Ciprofloxacin 500mg as a single dose (age 5–12y—250mg; <5y—30mg/kg—maximum 125mg), *or*
- Rifampicin 600mg bd for 2d (child 10mg/kg bd for 2d unless <1y when dose is 5mg/kg bd for 2d). Rifampicin colours urine red

Vaccination Routine childhood vaccination—➔ p. 619. In addition, offer unvaccinated close contacts of patients with confirmed meningococcal disease vaccination appropriate to the serotype of the confirmed case. Meningococcal vaccination (types A and C) may also be advised for travel to high-risk areas.

Category	High-risk criteria	Moderate- to high-risk criteria	Low-risk criteria
Behaviour	No response to social cues Appears ill to a healthcare professional Does not wake or, if roused, does not stay awake Weak high-pitched or continuous cry	Not responding normally to social cues No smile Wakes only with prolonged stimulation ↓ activity Parent/carer concern that the child is behaving differently from usual	Responds normally to social cues Content or smiles Stays awake or wakens quickly Strong normal cry or not crying
Respiratory	Grunting Apnoea O_2 saturation of <90% in air or ↑ O_2 requirement from baseline ↑ respiratory rate: <1y ≥60 breaths/min 1–2y ≥50 breaths/min 3–4y ≥40 breaths/min	O_2 saturation of <92% in air or ↑ O_2 requirement from baseline Nasal flaring ↑ respiratory rate: <1y 50–59 breaths/min 1–2y 40–49 breaths/min 3–4y 35–39 breaths/min	No high-risk or moderate- to high-risk criteria met
Circulation/ hydration	Heart rate <60bpm ↑ heart rate: <1y ≥160bpm 1–2y ≥150bpm 3–4y ≥140bpm	Capillary refill time ≥3s ↑ heart rate: <1y 150–159bpm 1–2y 140–149bpm 3–4y 130–139bpm	No high-risk or moderate- to high-risk criteria met
Temperature	Temperature <36°C If age <3mo, fever ≥38°C	If age 3–6mo, fever ≥39°C	Normal colour No high-risk or moderate- to high-risk criteria met
Skin and other	Mottled or ashen appearance Cyanosis of skin, lips, or tongue Non-blanching skin rash	Leg pain Cold hands/feet	

Figure 29.13 Risk stratification tool for children aged <5y with suspected sepsis[N]

Reproduced with permission from NICE (2016, updated 2017) Sepsis: recognition, diagnosis and early management. www.nice.org.uk/guidance/ng51

Late-onset effects Be alert for:

- ↓ hearing (audiology testing should be organized by the treating hospital)
- Ongoing neurological problems, e.g. fits, hemiparesis
- Orthopaedic problems, e.g. bone and joint damage, poor limb growth
- Psychosocial effects, e.g. learning disability, behavioural difficulties

Further information

NICE (2010, 2015) Meningitis (bacterial) and meningococcal septicaemia in under 16s: recognition, diagnosis and management. 🖱 www.nice.org.uk/guidance/cg102

NICE (2016, updated 2017) Sepsis: recognition, diagnosis and early management. 🖱 www.nice.org.uk/guidance/ng51

Telephone helplines for families

Meningitis Research Foundation ☎ 0808 8003344 🖱 www.meningitis.org
Meningitis Now ☎ 0808 8010338 🖱 www.meningitis-trust.org

Haemorrhage and shock

Shock Results from inadequate blood flow to the peripheral circulation. Signs include ↓ BP (± tachycardia), peripheral cyanosis, and ↓ urinary output. Treatment varies according to the underlying cause:

- **Anaphylactic shock** ➔ p. 1054
- **Septic shock** ➔ p. 1056
- **Hypovolaemic shock**
- **Cardiogenic shock** Inability of the heart to maintain sufficient blood flow (e.g. due to MI, arrhythmia, tamponade)—➔ p. 1065
- **Other rarer causes** Neurogenic (due to cerebral trauma or haemorrhage, e.g. head injury, subarachnoid haemorrhage); poisoning; liver failure

Hypovolaemic shock Occurs with insufficient circulating volume. *Signs*:

- **Initially** Tachycardia (pulse >100bpm), pallor, sweating ± restlessness
- **Later** Decompensation—sudden ↓ in pulse rate and BP. Young people may decompensate very rapidly—if tachycardic treat as a medical emergency—speed could be lifesaving

Causes Haemorrhage (internal or external) is the most usual cause:

- Gastrointestinal bleeding
- Nose bleeds—➔ p. 1062
- Ruptured thoracic/abdominal aneurysm
- Ruptured spleen ➔ p. 1062
- Gynaecological bleeding—very heavy period (➔ p. 684); miscarriage or ectopic pregnancy (➔ p. 767, p. 768)
- Bleeding associated with pregnancy—antepartum haemorrhage (➔ p. 810); postpartum haemorrhage (➔ p. 811)
- Bleeding associated with surgery, lacerations, or wounds—➔ p. 1062, p. 1062

Management Involves resuscitation with fluid replacement and treatment of the underlying cause.

Bleeding in terminally ill patients ➔ p. 1062

Ruptured aneurysm

Ruptured abdominal aortic aneurysm (AAA) In the community, death rate from ruptured AAA is ~90%. Consider ruptured AAA in any patient with ↓ BP and either back pain and/or atypical abdominal symptoms—especially if a pulsatile mass is palpable in the abdomen.

⚠ If known AAA, abdominal/back pain represents a ruptured AAA unless proved otherwise.

Dissecting thoracic aneurysm Typically presents with sudden tearing chest pain radiating to the back. Consider if ↓ BP and chest pain. As dissection progresses, branches of the aorta are sequentially occluded causing: hemiplegia (carotid artery); unequal pulses and BP in the two arms (subclavian artery); paraplegia (spinal arteries); acute renal failure (renal arteries); aortic incompetence (proximal extension); and MI (carotid arteries).

Management Lie the patient flat, and raise legs above waist height. Call for emergency ambulance assistance. If possible and equipment is available:

- Gain IV access and take blood for FBC and cross-matching—try to insert 2 large-bore cannulae; start IV fluids—give rapidly over 10–15min
- Give 100% oxygen (15L/min) (unless hypercapnic respiratory failure when give 24% oxygen)

Gastrointestinal bleeding Take all GI bleeding seriously; 7% of patients admitted to hospital with GI bleeding die. *Causes*: Table 29.2. *Presentation*:

- **Bleeding per rectum** Brisk bleeding is a medical emergency but often patients complain of small amounts of bleeding related to passage of stool. Mixed blood and stool implies bleeding proximal to the sigmoid colon; blood around the stool implies a more distal bleed; blood on the toilet paper or in the pan not mixed with stool is often from anal bleeding due to haemorrhoids or an anal fissure. ❗ Very heavy upper GI bleeds can present with fresh red bleeding PR
- **Haematemesis** Vomiting of blood—usually from upper GI bleeding but can rarely be swallowed blood from a nose bleed. Consider oesophageal varices if chronic liver disease and/or history of alcohol misuse
- **Melaena** Passage of black, offensive tarry stool consisting of altered blood. Usually indicates upper GI bleeding. ❗ Iron tablets turn the stool black
- **Coffee ground vomit** Vomiting of altered blood—looks like coffee granules. Implies upper GI bleeding—although less severe than fresh red blood
- **Other features** Faintness/dizziness may precede bleeding; if hypovolaemic shock: cold/clammy; ↑ respiratory rate; peripherally shut down; tachycardia; ↓ BP (or postural drop in BP)

Initial management Briefly assess the severity of the bleed from history alone (if telephone contact) ± examination. If a significant GI bleed is suspected, arrange immediate transfer to hospital via emergency ambulance. While awaiting transfer, ensure the patient is lying flat with legs above waist height (e.g. raised on a pillow). If with the patient and equipment is available:

- Insert a large-bore IV cannula—the opportunity may be lost by the time the ambulance crew arrives. If possible take a sample for FBC and cross-match on insertion. Start IV fluids
- Give 100% oxygen (15L/min) (↓ flow rate if hypercapnic respiratory failure)

Follow-up for patients with less severe bleeds Except for patients with haemorrhoids/anal fissure, all patients presenting with GI bleeding, even if the bleeding does not cause any circulatory compromise, require further investigation to establish the cause of the bleeding[N]:

- **Refer urgently to a lower GI team (to be seen in <2wk)** If abdominal/rectal mass; unexplained rectal bleeding aged ≥50y; or unexplained rectal bleeding aged <50y associated with abdominal pain, change in bowel habit, weight ↓, or iron deficiency anaemia
- **Refer urgently to an upper GI team (to be seen in <2wk)** If abdominal mass, dysphagia, or aged ≥55y and weight ↓, upper abdominal pain, reflux, and/or dyspepsia—otherwise, if haematemesis/coffee ground vomit and no circulatory compromise, refer routinely for upper GI endoscopy

Table 29.2 Causes of GI bleeding

General	Upper GI	Lower GI
Drugs—steroids, anticoagulants, NSAIDs Angiodysplasia (AV malformations are common) Haemangioma Bleeding disorders	Peptic ulcer Gastritis Mallory–Weiss tear Oesophagitis Oesophageal/gastric cancer Oesophageal varices	Diverticulitis Colitis—infectious or inflammatory Large bowel tumour or polyp Haemorrhoids Anal fissure

Wounds and lacerations Most patients with significant lacerations present directly to A&E. If a patient presents to general practice, perform immediate care (elevate bleeding limb and apply pressure to arrest bleeding). Advise nil by mouth and transfer to A&E by emergency ambulance.

Minor lacerations That can be managed in primary care ➲ p. 1089

Tetanus vaccination and prophylaxis ➲ p. 1089

Ruptured spleen May occur immediately following trauma or present days/weeks later. Diseased spleens (e.g. glandular fever, malaria, leukaemia) rupture more easily—advise to avoid contact sports/heavy lifting for >1mo after recovery.

Presentation

- History of abdominal trauma (may be trivial if diseased spleen)
- Blood loss: ↑ respiratory rate, tachycardia, ↓ BP ± postural drop, pallor
- Peritoneal irritation: guarding, abdominal rigidity, shoulder tip pain
- Paralytic ileus: abdominal distension, lack of bowel sounds

⚠ *Action* If suspected, admit as a blue light surgical emergency.

Postoperative haemorrhage Occasionally patients may call for GP assistance after discharge following a surgical procedure. Consider postoperative haemorrhage if any symptoms/signs of shock, particularly if recent discharge and short hospital stay (e.g. day case procedure). If suspected, readmit to hospital for assessment by emergency ambulance.

⚠ Tonsillectomy carries a small risk of severe haemorrhage. Readmit any patient with bleeding postoperatively for observation.

Nose bleeds (epistaxis) Usually due to ruptured blood vessels on the nasal septum. *Causes*:

- Young Nose picking, coryza, allergic rhinitis, blood dyscrasias
- Elderly Degenerative arterial disease, ↑ BP, nose picking, coryza, allergic rhinitis, medication (anticoagulants or aspirin), blood dyscrasias, telangiectasia, tumour. Often no cause is found

Management Figure 29.14

Recurrent minor nosebleeds Refer to ENT for examination and electrocautery.

Bleeding in terminal illness If severe, life-threatening bleed, make a decision whether the cause of the bleed is treatable or a terminal event. This is best done in advance, but bleeding cannot always be predicted.

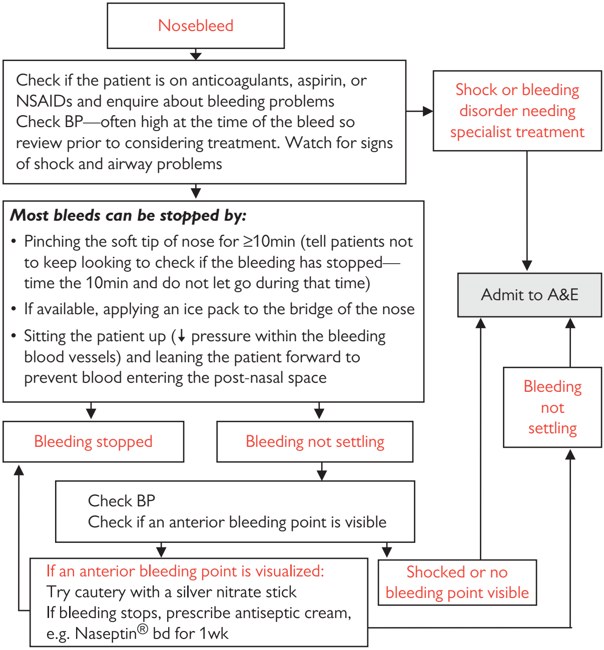
Active treatment Call for an emergency ambulance; lie the patient flat with feet raised; treat underlying cause if possible. If equipment is available:

- Gain IV access and resuscitate with IV fluids given rapidly over 10–15min
- Give 100% oxygen (15L/min) (24% if hypercapnic respiratory failure)

❗ If bleeding is from a lung tumour, protect the airway and lie the patient on the side of the tumour. Unless a patient is very near to death, admit all palliative patients with non-life-threatening GI bleeds related to their underlying condition. Palliative treatment options include laser treatment and arterial embolization—both can be performed on frail patients.

If no active treatment is indicated Stay with the patient:

- Give sedative medication, e.g. midazolam 20–40mg sc or IV, or diazepam 10–20mg PR ± analgesia
- Support carers, as big bleeds are extremely distressing


Nosebleed
Shock or bleeding disorder needing specialist treatment
Admit to A&E
Bleeding not settling
Bleeding stopped
Bleeding not settling
Shocked or no bleeding point visible

Check if the patient is on anticoagulants, aspirin, or NSAIDs and enquire about bleeding problems
Check BP—often high at the time of the bleed so review prior to considering treatment. Watch for signs of shock and airway problems

Most bleeds can be stopped by:

- Pinching the soft tip of nose for ≥10min (tell patients not to keep looking to check if the bleeding has stopped—time the 10min and do not let go during that time)
- If available, applying an ice pack to the bridge of the nose
- Sitting the patient up (↓ pressure within the bleeding blood vessels) and leaning the patient forward to prevent blood entering the post-nasal space

Check BP
Check if an anterior bleeding point is visible

If an anterior bleeding point is visualized:
Try cautery with a silver nitrate stick
If bleeding stops, prescribe antiseptic cream, e.g. Naseptin® bd for 1wk

Figure 29.14 Acute management of nosebleed in the community

Further information

NICE (2015, updated 2017) Suspected cancer: recognition and referral.
www.nice.org.uk/guidance/ng12

Chest pain and palpitations

Chest pain Common symptom.

⚠ Always think—could this be an acute coronary syndrome (ACS), PE, dissecting aneurysm, or pericarditis?

On receiving the call for assistance Ask:

- Nature and location of the pain
- Duration of the pain
- Other associated symptoms—sweating, nausea, shortness of breath, palpitations
- Past medical history (particularly heart disease, high cholesterol)
- Family history (particularly heart disease)
- Smoker?

Action

- Consider differential diagnosis (Table 29.3)
- If ACS is suspected call for ambulance assistance before (or instead of) visiting
- Otherwise visit (or arrange surgery appointment), assess, and treat according to cause

History Ask about:

- Site and nature of pain. Any history of trauma?
- Duration
- Associated symptoms (e.g. breathlessness, nausea)
- Provoking and relieving factors
- PMH, FH (e.g. heart disease), drug history, smoking history

Examination

- Check BP in both arms
- General appearance—distress, sweating, pallor
- JVP and carotid pulse
- Respiratory rate
- Apex beat
- Heart sounds
- Lung fields
- Local tenderness
- Pain on movement of chest
- Skin rashes
- Swelling or tenderness of legs (?DVT)

Investigations ECG and CXR may be helpful.

Palpitations Uncomfortable awareness of heart beat. Can be physiological (e.g. after exercise, at times of stress) or signify arrhythmia. Can cause a feeling of faintness or even collapse (e.g. Stokes–Adams attack, due to AV block). Ask the patient to tap out the rhythm.

- Bradycardia ➲ p. 242
- Occasional missed beat Suggests ventricular ectopics—➲ p. 238
- Tachycardia ➲ p. 238

Table 29.3 Causes of acute chest pain

Diagnosis	Features
Acute coronary syndrome ➲ p. 1066	Band-like chest pain around the chest or central chest pressure/ dull ache ± radiation to shoulders, arms (L > R), neck, and/or jaw Often associated with nausea, sweating, and/or shortness of breath
Pericarditis ➲ p. 247	Sharp, constant sternal pain relieved by sitting forward May radiate to left shoulder ± arm or into the abdomen Worse lying on the left side and on inspiration, swallowing, and coughing
Dissecting thoracic aneurysm ➲ p. 1060	Typically presents with sudden tearing chest pain radiating to the back Consider in any patient with chest pain (especially if radiates through to the back) and ↓ BP
PE ➲ p. 1070	Acute dyspnoea, sharp chest pain (worse on inspiration), haemoptysis, and/or syncope. Tachycardic and mild pyrexia
Pleurisy	Sharp, localized chest pain, worse on inspiration May be associated with symptoms and signs of a chest infection
Pneumothorax ➲ p. 1070	Sudden onset of pleuritic chest pain and/or ↑ breathlessness ± pallor and tachycardia
Oesophageal spasm/ oesophagitis	Central chest pain. May be associated with acid reflux (although not always) May be described as burning but often indistinguishable from cardiac pain May respond to antacids
Musculoskeletal pain	Localized pain—worse on movement May be a history of injury
Shingles	Intense, often sharp, unilateral pain Responds poorly to analgesia Pain may be present several days before rash appears
Costochondritis	Inflammation of the costochondral junctions—tenderness over the costochondral junction and pain in the affected area on springing the chest wall
Bornholm's disease	Unilateral chest and/or abdominal pain, rhinitis. Coxsackie virus infection Treat with simple analgesia
Idiopathic chest pain	No cause apparent. Common Affects young people > elderly people. ♀ > ♂

⚠ If a patient is acutely unwell with chest pain and the cause is not clear, err on the side of caution and admit for further assessment.

Cardiogenic shock (e.g. due to MI, arrhythmia, tamponade). *Signs*:

- Hypotension—systolic BP <80–90mmHg
- Pulse rate may be normal, ↑, or ↓

Action Sit the patient up if possible. Call for ambulance assistance. If equipment is available:

- Gain IV access. Treat underlying cause, e.g. atropine for bradycardia; diamorphine, furosemide, and GTN spray (if tolerated) for acute LVF
- Give 100% oxygen (15L/min) (↓ flow rate if hypercapnic respiratory failure)

Acute coronary syndrome

The term acute coronary syndrome (ACS) covers:

- **Myocardial infarction** Both ST segment elevation MI (STEMI) and non-ST segment elevation MI (NSTEMI), *and*
- **Unstable angina**

Initial primary care management is the same for STEMI, NSTEMI, and unstable angina.

Presentation May be new onset or a rapid deterioration of stable angina. Presenting features include:

- Sustained central chest pain (>15min)—typically described as central crushing/pressure, band-like pain
- Pain radiating to the arms, jaw, back, or upper abdomen (may be the only symptom)
- Symptoms resulting from sympathetic autonomic stimulation, e.g. nausea, vomiting, sweating
- Symptoms relating to cardiogenic shock (➔ p. 1065), e.g. breathlessness, hypotension, collapse

Other factors to consider

- Does the patient have risk factors for cardiac disease?
- Has the patient had previous investigations for chest pain? If so, what investigations were done, when, and what were the results?
- Does the patient have a history of ischaemic heart disease? If so, what is the current treatment and what has been tried in the past?

⚠ Diagnosis of ACS is sometimes difficult (e.g. patients with DM may have silent MI): have a high index of suspicion.

Examination Pulse, BP, JVP, heart sounds, chest (?pulmonary oedema).

ECG Do not do an ECG if it delays transfer of the patient to hospital. Normal ECG does not exclude ACS. If an ECG is done, send it electronically to the receiving hospital or send a copy of the ECG with the patient to hospital. ECG changes—Table 29.4.

Table 29.4 Features of STEMI, NSTEMI, and unstable angina

	STEMI	NSTEMI	Unstable angina
Chest pain?	Yes	Yes	Yes
ECG changes?	ST elevation (≥1mm in ≥2 adjacent limb leads or ≥2mm in ≥2 adjacent anterior chest leads) *or* new LBBB	Normal or signs of myocardial ischaemia: • ST-segment depression • T-wave inversion/flattening	Normal or signs of myocardial ischaemia: • ST-segment depression • T-wave inversion/flattening
Troponin levels?[a]	Raised	Raised	Normal

[a] Usually done in hospital. Indicates myocardial muscle necrosis. Becomes +ve 3–6h after onset of pain and may remain +ve for 7–14d.

⚠ Action

When the call for assistance is made If ACS is suspected, arrange immediate transfer to hospital. For reperfusion interventions (thrombolysis or percutaneous coronary intervention) to be effective, they must be carried out as soon as possible after the onset of pain. Seeing the patient before arranging transfer introduces unnecessary delays.

If attending the patient (e.g. patient reports chest pain during a same day GP surgery appointment). Call for an emergency ambulance to transfer the patient to hospital then:

- Give pain relief with either sublingual GTN or IV/IM opioid (e.g. morphine 5–10mg—half dose if elderly/frail) or both
- Give aspirin 300mg po (unless contraindicated)
- Consider giving IV/IM antiemetic (e.g. metoclopramide 10mg)
- Measure peripheral oxygen saturation with pulse oximeter—only give oxygen if saturations are <94%. Aim for saturations of 94–98% (88–92% if known hypercapnic respiratory failure and at risk of CO_2 retention)
- If bradycardia, consider giving atropine 500 micrograms IV and further doses of 500 micrograms if needed to a maximum of 3mg

❗ To avoid inadvertent replication of medication when the patient reaches hospital, record all drugs that have been given (name of the drug, dose and route of administration, time administered). Send this information to hospital with the patient.

Thrombolysis in general practice May be appropriate in places where transfer to hospital takes >30min. Special training and equipment are needed.

Late calls If the patient is seen pain-free after an acute episode.

Admit as an emergency If the patient

- Has signs of complications that require emergency admission (e.g. pulmonary oedema)
- Is seen <12h after the acute episode and the ECG is abnormal or ECG is unavailable

Arrange same-day specialist assessment If the patient:

- Is seen <12h after the acute episode, is well, and ECG is normal
- Is seen 12–72h after the acute episode and there are no reasons for immediate emergency admission

If the patient is seen >72h after an acute episode and is well Assess clinically, perform ECG, and arrange blood test for troponin. Refer to cardiology for routine follow-up/more urgently, depending on the clinical situation. For patients with new-onset chest pain, most areas run rapid access chest pain clinics.

In the interim, start regular aspirin; supply with GTN spray, and warn to call for assistance (by calling ambulance and/or emergency GP) if chest pain lasts >15min despite GTN spray.

Further information

NICE (2010, updated 2016) Chest pain of recent onset. 🖱 www.nice.org.uk/guidance/cg95

Acute breathlessness in adults

⚠ Attend as soon as possible after receiving the call for help. If there is likely to be any delay, call for emergency ambulance assistance.

On arrival

- Be calm and reassuring—breathlessness is frightening and panic only adds to the sensation of being breathless
- Direct history and examination to finding the cause as quickly as possible—treat according to the cause
- If no cause can be found—do not delay—admit to hospital as an acute medical emergency

Causes Table 29.5

Acute heart failure Severe acute breathlessness due to pulmonary oedema. Urgent action is needed to save life.

Presenting features

- Sudden acute breathlessness
- Fatigue
- Cough ± haemoptysis (usually pink and frothy)
- Tends to occur at night
- Some relief gained from sitting/standing

Signs

- Dyspnoea, ↑ respiratory rate ± ↓ peripheral oxygen saturations
- Tachycardia—gallop rhythm may be present
- Coarse wet-sounding crackles at both bases
- Ankle/sacral oedema if right heart failure also present
- ± hypotension

Action If severe, call for emergency ambulance support; sit the patient up; be reassuring—it is frightening to be very short of breath. If available:

- Give oxygen—aim for peripheral O_2 saturations of 94–98% (if history of hypercapnic respiratory failure, aim for saturations of 88–92%)
- Consider po or slow IV furosemide 20–60mg (or bumetanide 1–2mg)

Admission Depends on severity and cause of attack, response to treatment, and social support. Always admit if:

- Alone at home and/or inadequate social support
- Suspected cause of acute LVF warrants admission (e.g. acute MI)
- Very breathless and no improvement over 30min with treatment at home
- Hypotension or arrhythmia

If new suspected heart failure and not admitting

- Check FBC, renal function, and serum natriuretic peptides (heart failure is ruled out if B-type natriuretic peptide (BNP) <100ng/L or N-terminal pro-B-type natriuretic peptide (NT-proBNP) < 300ng/L)
- Once stabilized, start ACE inhibitor and β-blocker (unless heart rate <50bpm or 2nd/3rd-degree heart block)
- Refer for specialist assessment and echocardiography (e.g. cardiac breathlessness clinic) to be seen in <2wk

Table 29.5 Causes of acute breathlessness

Diagnosis	Features
Asthma ➔ p. 1072 (adult); ➔ p. 1075 (child)	Breathlessness and wheeze. Usually in association with a past history of asthma, although can present *de novo* Signs of a severe attack include: inability to speak in sentences, tachycardia, pulsus paradoxus, ↑ respiratory rate, use of accessory muscles of respiration, ↓ peripheral O_2 saturation, drowsiness, or exhaustion
Anaphylaxis ➔ p. 1054	Often history of anaphylaxis/severe allergic reaction. Sudden onset/rapid progression of symptoms (minutes) and life-threatening: ● Airway problems (difficulty breathing, stridor) ● Breathing problems (↑ respiratory rate, wheeze, ↓ peripheral O_2 saturation); and/or ● Circulation problems (↑ pulse rate, ↓ BP, shock, ↓ consciousness) ± skin changes (e.g. urticaria, flushing, angio-oedema, rhinitis)
Acute heart failure ➔ p. 1066	**Symptoms** Sudden acute breathlessness Fatigue Cough ± haemoptysis Tends to occur at night Some relief from sitting/standing **Signs** Dyspnoea Tachycardia ± gallop rhythm Coarse crackles at both bases Ankle/sacral oedema ± hypotension
Arrhythmia ➔ p. 238	Usually palpitations (although not always). Associated with chest pain, collapse or funny turns, sweating, breathlessness, and/or hyperventilation. May be a PMH/FH of similar symptoms or thyroid disease
PE ➔ p. 1070	Acute dyspnoea, sharp chest pain (worse on inspiration), haemoptysis, and/or syncope. Tachycardic and mild pyrexia
Acute exacerbation of COPD ➔ p. 288	Worsening of previously stable COPD. Presents with ≥1 of: ↑ dyspnoea ↓ exercise tolerance or ↑ fatigue ↑ fluid retention ↑ wheeze/chest tightness ↑ cough ↑ sputum purulence or volume Upper airways symptoms, e.g. rhinorrhoea, sore throat New-onset cyanosis Acute confusion
Pneumonia ➔ p. 294	Breathlessness, cough, fever, sputum ± sharp, localized chest pain, worse on inspiration
Viral respiratory distress syndrome ➔ p. 299	History of viral infection – usually dry cough and fever. Rapidly worsening breathlessness, low peripheral oxygen saturation, and few chest signs. Admit as an acute medical emergency
Pneumothorax ➔ p. 1070	Sudden onset of pleuritic chest pain and/or ↑ breathlessness ± pallor and tachycardia
Choking ➔ p. 1048 (adult); ➔ p. 1050 (child)	Think of foreign body airway obstruction if history of sudden onset of stridor or symptoms of respiratory distress
SVC obstruction ➔ p. 1024	Acute breathlessness, headache worse on stooping, swelling of the face and/or neck with fixed elevation of JVP—admit for assessment
Air hunger due to shock ➔ p. 1060	Inadequate blood flow to the peripheral circulation—usually associated with ↓ BP (± tachycardia) and peripheral cyanosis
Hyperventilation ➔ p. 1081	Breathlessness associated with fear, terror, and a sense of impending doom

Further information

NICE (2014) Acute heart failure: diagnosis and management. 🖱 www.nice.org.uk/guidance/cg187

Pulmonary embolism and pneumothorax

Pulmonary embolism Venous thrombi—usually from a deep vein thrombosis in the leg—pass into the pulmonary circulation and block blood flow to the lungs. Without treatment 20% with proximal deep vein thrombosis develop pulmonary embolus (PE). PE is listed as a cause of death on 12,000–13,000 death certificates every year in the UK.

Risk factors

- Immobility—long flight or bus journey, post-op, plaster cast
- Smoking
- Malignancy
- Combined hormonal contraception
- Pregnancy or puerperium
- Past history or family history of DVT, PE, or clotting tendency

Presentation Have a high level of suspicion. Consider PE if: acute dyspnoea, pleuritic chest pain, haemoptysis, syncope. Look for a source of emboli—although DVT may not be clinically obvious. Signs include: ↑ respiratory rate, ↑ pulse rate, ↓ BP, ↓ peripheral oxygen saturations ± cyanosis, ↑ JVP, pleural rub.

Differential diagnosis

- Pneumonia and pleurisy
- Acute coronary syndrome
- Other causes of acute breathlessness—acute heart failure, asthma, exacerbation of COPD, pneumothorax, shock (e.g. due to anaphylaxis), arrhythmia, hyperventilation
- Other causes of acute chest pain—aortic dissection, rib fracture, musculoskeletal chest pain, pericarditis, oesophageal spasm, shingles

Immediate action if severe symptoms Large clots can be rapidly fatal. Call for emergency ambulance support. Give oxygen as soon as possible (aim to keep peripheral oxygen saturations at 94–98%) and admit as an acute medical emergency.

If stable but suspected diagnosis of PE[N] Do a Wells PE score (Table 29.6):

- **If PE is likely** Refer to the acute medical team for a CT pulmonary angiogram (CTPA). If CTPA cannot be carried out immediately, anticoagulate with sc heparin or a direct oral anticoagulant (DOAC) while awaiting CTPA. Consider proximal leg vein USS if CTPA is –ve and DVT is likely (➔ p. 260)
- **If PE is unlikely** Check urgent D-dimer blood test. If +ve, treat as if PE is likely. If –ve, reassure and look for an alternative diagnosis. If unable to check an urgent D-dimer, refer to the acute hospital medical team

Anticoagulation for confirmed PE If confirmed PE, patients are anticoagulated with either warfarin (*target INR*: 2.5) or a DOAC for ≥3mo (➔ p. 648).

Spontaneous pneumothorax Risk factors:

- Previous pneumothorax
- Smoking
- Ascent in an aeroplane
- Diving

Cause

- **In patients <40y** Usually due to rupture of a pleural bleb. The typical patient is tall, thin, and male (♂:♀ ≈6:1)
- **Patients >40y** Usually due to COPD (70–80%)
- **Rarer causes** Asthma, pneumonia, TB, lung cancer, pulmonary fibrosis

Table 29.6 Wells PE score

Clinical feature	Score
Clinical symptoms/signs of DVT	3
Alternative diagnosis is less likely than PE	3
Heart rate >100bpm	1.5
Immobilization for >3d or surgery <4wk previously	1.5
Previous DVT/PE	1.5
Haemoptysis	1
Malignancy (on treatment currently or in the past 6mo, or palliative)	1
PE is likely if score >4 *PE is unlikely if score ≤4*	

Source: data from Wells et al. *Thromb Haemost* 2000;**83**:358.

Presentation Sudden onset of pleuritic chest pain or ↑ breathlessness ± pallor and tachycardia. Look for resonant percussion note, ↓ or absent breath sounds—signs may be absent if the pneumothorax is small.

Management Refer for same-day CXR. If pneumothorax is confirmed, seek specialist advice about further management. Small pneumothoraces usually resolve spontaneously (50% collapse takes ~40d to resorb)—monitor until completely resolved. Larger pneumothoraces may require admission for aspiration or a chest drain. Smoking cessation ↓ risk of recurrence.

Traumatic pneumothorax Trauma may not initially be obvious—ask about injections around the chest area, e.g. acupuncture (to neck and shoulders as well as chest); aspiration of breast lump, etc. Presentation and management is as for spontaneous pneumothorax.

Tension pneumothorax Complication of traumatic pneumothorax; rare after spontaneous pneumothorax. A valvular mechanism develops—air is sucked into the pleural space during inspiration but cannot be expelled during expiration. The pressure within the pleural space ↑ and the lung deflates further; the mediastinum shifts to the opposite side of the chest, and venous return ↓. Can be rapidly fatal. *Clinical features*:

- Agitated and distressed patient, often with a history of chest trauma
- Tachycardia
- Sweating
- Signs of large pneumothorax—↓ breath sounds; ↓ chest movement on the affected side
- Mediastinal shift—trachea deviates away from the pneumothorax side

⚠ *Action* If tension pneumothorax is suspected, sit the patient upright if possible. Insert a large-bore cannula through the 2nd intercostal space of the chest wall in the mid-clavicular line on the side of the pneumothorax to relieve the pressure in the pleural space. Transfer as an emergency to hospital.

Further information

NICE (2012, updated 2015) Venous thromboembolic diseases. 🖱 www.nice.org.uk/guidance/cg144

Acute asthma in adults

Many deaths from asthma are preventable. Delay can be fatal. Factors leading to poor outcome include:

- Doctors failing to assess severity by objective measurement
- Patients or relatives failing to appreciate severity
- Underuse of corticosteroids

⚠ Regard each emergency asthma consultation as acute severe asthma until proven otherwise.

Risk factors for developing fatal or near-fatal asthma A combination of severe asthma recognized by ≥1 of:

- Previous near-fatal asthma
- Previous admission for asthma—especially if within 1y
- Requiring ≥3 classes of asthma medication
- Heavy use of β_2-agonist
- Repeated attendances at A&E for asthma care—especially if within 1y
- Brittle asthma

And adverse behavioural or psychosocial features recognized by ≥1 of:

- Non-compliance with treatment or monitoring
- Failure to attend appointments
- Self-discharge from hospital
- Psychosis, depression, other psychiatric illness, or deliberate self-harm
- Current or recent major tranquillizer use
- Denial
- Alcohol or drug misuse
- Obesity
- Learning difficulties
- Employment/income problems
- Social isolation
- Childhood abuse
- Severe marital/legal/domestic stress

Assess and record

- Symptoms and response to self-treatment
- Respiratory rate, amount of wheeze, and level of breathlessness
- Peak expiratory flow rate (PEFR)
- Peripheral oxygen saturation (SpO_2)
- Pulse rate and BP—↑ heart rate generally reflects ↑ severity
- Degree of agitation and conscious level

Severity of an acute asthma exacerbation

⚠ Patients with severe or life-threatening attacks may not be distressed and may not have all the characteristic abnormalities of severe asthma. The presence of any should alert the treating healthcare professional.

Moderate asthma exacerbation

- Increasing symptoms
- No features of acute, severe asthma
- PEFR >50–75% predicted

Acute severe asthma Any one of:

- PEFR 33–50% best or predicted
- Respiratory rate ≥25 breaths/min
- Heart rate ≥110/min
- Inability to complete sentences in 1 breath

⚠ *Life-threatening asthma* Any 1 of the following with severe asthma:

- PEFR <33% best/predicted
- O_2 saturation <92%
- Silent chest
- Cyanosis
- Feeble respiratory effort
- Bradycardia
- Dysrhythmia
- Hypotension
- Exhaustion
- Confusion
- Coma

Near-fatal asthma Respiratory acidosis and/or requiring mechanical ventilation with ↑ inflation pressures.

Brittle asthma

- **Type 1** Wide PEFR variability (>40% diurnal variation for >50% of the time for a period of >150d) despite intense therapy
- **Type 2** Sudden severe attacks on a background of apparently well-controlled asthma

⚠ Management

See Figure 29.15, ➔ p. 1074.

Admit to hospital as an acute medical emergency if

- Life-threatening features
- Features of acute severe asthma present after initial treatment
- Previous near-fatal asthma

Lower threshold for admission if

- Afternoon or evening attack
- Recent nocturnal symptoms or hospital admission
- Previous severe attacks
- Patient unable to assess own condition
- Concern over social circumstances

If admitting the patient to hospital

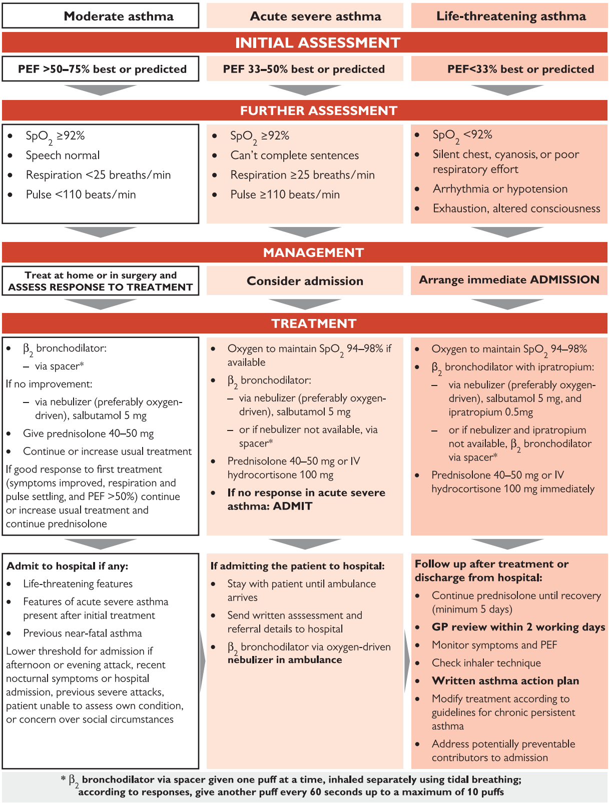
- Stay with the patient until the ambulance arrives
- Send written assessment including NEWS2 score (➔ p. 1036) and treatment given to the hospital
- Give β_2 bronchodilator via an oxygen-driven nebulizer in the ambulance

Follow-up after treatment or discharge from hospital

- Arrange review in primary care within 48h
- Monitor symptoms and PEFR
- Check inhaler technique
- Provide a written asthma action plan
- Modify treatment according to guidelines for chronic persistent asthma
- Address potentially preventable contributors to admission

Further information

BTS/SIGN (2019) British guideline on the management of asthma. 🖱 www.sign.ac.uk/sign-158-british-guideline-on-the-management-of-asthma.html



Moderate asthma	**Acute severe asthma**	**Life-threatening asthma**
INITIAL ASSESSMENT		
PEF >50–75% best or predicted	**PEF 33–50% best or predicted**	**PEF<33% best or predicted**
FURTHER ASSESSMENT		
• SpO_2 ≥92% • Speech normal • Respiration <25 breaths/min • Pulse <110 beats/min	• SpO_2 ≥92% • Can't complete sentences • Respiration ≥25 breaths/min • Pulse ≥110 beats/min	• SpO_2 <92% • Silent chest, cyanosis, or poor respiratory effort • Arrhythmia or hypotension • Exhaustion, altered consciousness
MANAGEMENT		
Treat at home or in surgery and ASSESS RESPONSE TO TREATMENT	**Consider admission**	**Arrange immediate ADMISSION**
TREATMENT		
• β_2 bronchodilator: – via spacer* If no improvement: – via nebulizer (preferably oxygen-driven), salbutamol 5 mg • Give prednisolone 40–50 mg • Continue or increase usual treatment If good response to first treatment (symptoms improved, respiration and pulse settling, and PEF >50%) continue or increase usual treatment and continue prednisolone	• Oxygen to maintain SpO_2 94–98% if available • β_2 bronchodilator: – via nebulizer (preferably oxygen-driven), salbutamol 5 mg – or if nebulizer not available, via spacer* • Prednisolone 40–50 mg or IV hydrocortisone 100 mg • **If no response in acute severe asthma: ADMIT**	• Oxygen to maintain SpO_2 94–98% • β_2 bronchodilator with ipratropium: – via nebulizer (preferably oxygen-driven), salbutamol 5 mg, and ipratropium 0.5mg – or if nebulizer and ipratropium not available, β_2 bronchodilator via spacer* • Prednisolone 40–50 mg or IV hydrocortisone 100 mg immediately
Admit to hospital if any: • Life-threatening features • Features of acute severe asthma present after initial treatment • Previous near-fatal asthma Lower threshold for admission if afternoon or evening attack, recent nocturnal symptoms or hospital admission, previous severe attacks, patient unable to assess own condition, or concern over social circumstances	**If admitting the patient to hospital:** • Stay with patient until ambulance arrives • Send written asssessment and referral details to hospital • β_2 bronchodilator via oxygen-driven **nebulizer in ambulance**	**Follow up after treatment or discharge from hospital:** • Continue prednisolone until recovery (minimum 5 days) • **GP review within 2 working days** • Monitor symptoms and PEF • Check inhaler technique • **Written asthma action plan** • Modify treatment according to guidelines for chronic persistent asthma • Address potentially preventable contributors to admission

*** β_2 bronchodilator via spacer given one puff at a time, inhaled separately using tidal breathing; according to responses, give another puff every 60 seconds up to a maximum of 10 puffs**

Figure 29.15 Management of acute severe asthma in adults[G]

This figure is reproduced from *SIGN 158: British guideline on the management of asthma*, by kind permission of the Scottish Intercollegiate Guidelines Network.

Acute asthma in children

Assess and record

- Pulse rate—↑ heart rate generally reflects ↑ severity
- Respiratory rate, breathlessness, and pulse oximetry (if available)
- Use of accessory muscles—best noted by palpation of neck muscles
- Amount of wheezing
- Degree of agitation and conscious level

Levels of severity ❗ If a patient has signs and symptoms across categories, always treat according to the most severe features.

- Child >5y See Figure 29.16, ➔ p. 1076
- Child 2–5y See Figure 29.17, ➔ p. 1077

Child <2y Assessment of children <2y can be difficult.

- Moderate asthma SpO$_2$ ≥92%; still feeding; audible wheezing; using accessory muscles
- Acute severe asthma SpO$_2$ <92%; too breathless to feed; marked respiratory distress; cyanosis
- Life-threatening asthma Poor respiratory effort; apnoea; cyanosis

Management

- Child >5y See Figure 29.16, ➔ p. 1076
- Child 2–5y See Figure 29.17, ➔ p. 1077

Child <2y Intermittent wheezing attacks are usually in response to viral infection, and response to bronchodilators is inconsistent.

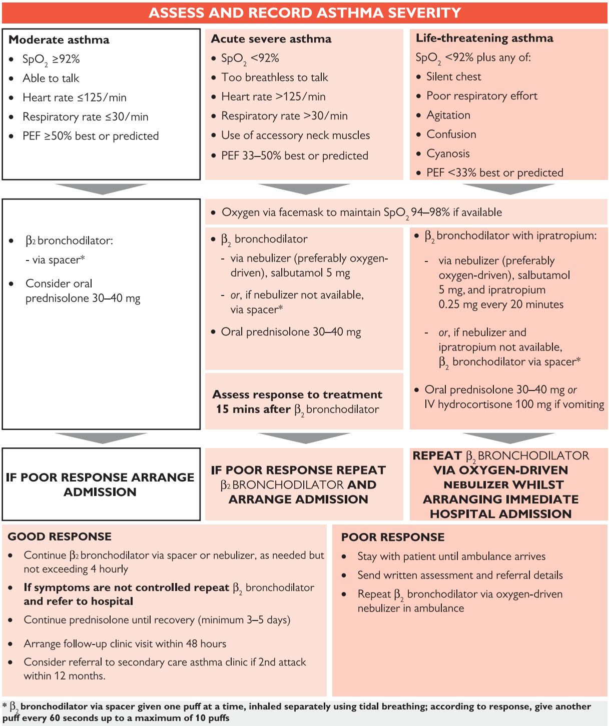
- If mild/moderate wheeze Consider a trial of a bronchodilator if symptoms are of concern—use a metered dose inhaler and spacer with a face mask. If positive response to bronchodilator, consider giving prednisolone 10mg po for 3d. If no response consider alternative diagnosis (aspiration pneumonitis, pneumonia, bronchiolitis, tracheomalacia, CF, congenital anomaly) and/or admit
- If severe wheezing Admit to hospital
- If any life-threatening features Admit immediately as a blue light emergency

Follow-up after treatment or discharge from hospital

- GP review within 1wk
- Monitor symptoms, PEFR, and check inhaler technique
- Provide a written asthma action plan
- Modify treatment according to guidelines for chronic persistent asthma
- Address potentially preventable contributors to admission

Further information

BTS/SIGN (2019) British guideline on the management of asthma 🖱 www.sign.ac.uk/sign-158-british-guideline-on-the-management-of-asthma.html



ASSESS AND RECORD ASTHMA SEVERITY

Moderate asthma
- SpO_2 ≥92%
- Able to talk
- Heart rate ≤125/min
- Respiratory rate ≤30/min
- PEF ≥50% best or predicted

Acute severe asthma
- SpO_2 <92%
- Too breathless to talk
- Heart rate >125/min
- Respiratory rate >30/min
- Use of accessory neck muscles
- PEF 33–50% best or predicted

Life-threatening asthma

SpO_2 <92% plus any of:
- Silent chest
- Poor respiratory effort
- Agitation
- Confusion
- Cyanosis
- PEF <33% best or predicted

Moderate asthma:
- β_2 bronchodilator:
  - via spacer*
- Consider oral prednisolone 30–40 mg

Acute severe and life-threatening asthma:
- Oxygen via facemask to maintain SpO_2 94–98% if available

Acute severe asthma:
- β_2 bronchodilator
  - via nebulizer (preferably oxygen-driven), salbutamol 5 mg
  - *or*, if nebulizer not available, via spacer*
- Oral prednisolone 30–40 mg

Assess response to treatment 15 mins after β_2 bronchodilator

Life-threatening asthma:
- β_2 bronchodilator with ipratropium:
  - via nebulizer (preferably oxygen-driven), salbutamol 5 mg, and ipratropium 0.25 mg every 20 minutes
  - *or*, if nebulizer and ipratropium not available, β_2 bronchodilator via spacer*
- Oral prednisolone 30–40 mg *or* IV hydrocortisone 100 mg if vomiting

IF POOR RESPONSE ARRANGE ADMISSION

IF POOR RESPONSE REPEAT β_2 BRONCHODILATOR **AND ARRANGE ADMISSION**

REPEAT β_2 BRONCHODILATOR **VIA OXYGEN-DRIVEN NEBULIZER WHILST ARRANGING IMMEDIATE HOSPITAL ADMISSION**

GOOD RESPONSE
- Continue β_2 bronchodilator via spacer or nebulizer, as needed but not exceeding 4 hourly
- **If symptoms are not controlled repeat** β_2 bronchodilator **and refer to hospital**
- Continue prednisolone until recovery (minimum 3–5 days)
- Arrange follow-up clinic visit within 48 hours
- Consider referral to secondary care asthma clinic if 2nd attack within 12 months.

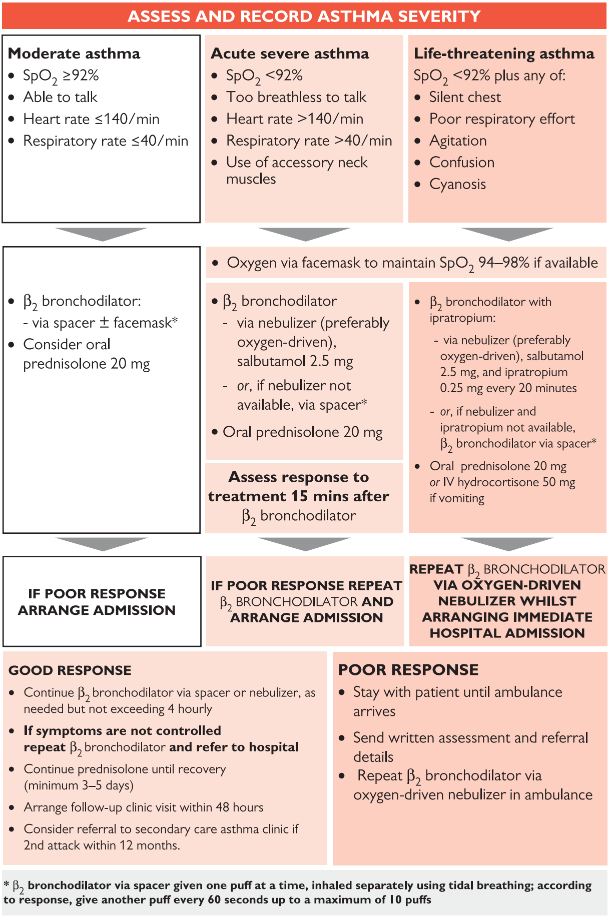
POOR RESPONSE
- Stay with patient until ambulance arrives
- Send written assessment and referral details
- Repeat β_2 bronchodilator via oxygen-driven nebulizer in ambulance

***** β_2 **bronchodilator via spacer given one puff at a time, inhaled separately using tidal breathing; according to response, give another puff every 60 seconds up to a maximum of 10 puffs**

Figure 29.16 Management of acute severe asthma in children aged >5y^{G}

This figure is reproduced from *SIGN 158: British guideline on the management of asthma*, by kind permission of the Scottish Intercollegiate Guidelines Network.

⚠ *Lower threshold for admission if*
- Attack in late afternoon or at night
- Recent hospital admission or previous severe attack
- Concern over social circumstances or ability to cope at home



ASSESS AND RECORD ASTHMA SEVERITY

Moderate asthma
- SpO_2 ≥92%
- Able to talk
- Heart rate ≤140/min
- Respiratory rate ≤40/min

Acute severe asthma
- SpO_2 <92%
- Too breathless to talk
- Heart rate >140/min
- Respiratory rate >40/min
- Use of accessory neck muscles

Life-threatening asthma

SpO_2 <92% plus any of:
- Silent chest
- Poor respiratory effort
- Agitation
- Confusion
- Cyanosis

Moderate asthma:
- β_2 bronchodilator:
  - via spacer ± facemask*
- Consider oral prednisolone 20 mg

Acute severe and life-threatening asthma:
- Oxygen via facemask to maintain SpO_2 94–98% if available

Acute severe asthma:
- β_2 bronchodilator
  - via nebulizer (preferably oxygen-driven), salbutamol 2.5 mg
  - *or*, if nebulizer not available, via spacer*
- Oral prednisolone 20 mg

Assess response to treatment 15 mins after β_2 bronchodilator

Life-threatening asthma:
- β_2 bronchodilator with ipratropium:
  - via nebulizer (preferably oxygen-driven), salbutamol 2.5 mg, and ipratropium 0.25 mg every 20 minutes
  - *or*, if nebulizer and ipratropium not available, β_2 bronchodilator via spacer*
- Oral prednisolone 20 mg *or* IV hydrocortisone 50 mg if vomiting

IF POOR RESPONSE ARRANGE ADMISSION

IF POOR RESPONSE REPEAT β_2 BRONCHODILATOR **AND ARRANGE ADMISSION**

REPEAT β_2 BRONCHODILATOR **VIA OXYGEN-DRIVEN NEBULIZER WHILST ARRANGING IMMEDIATE HOSPITAL ADMISSION**

GOOD RESPONSE
- Continue β_2 bronchodilator via spacer or nebulizer, as needed but not exceeding 4 hourly
- **If symptoms are not controlled repeat** β_2 bronchodilator **and refer to hospital**
- Continue prednisolone until recovery (minimum 3–5 days)
- Arrange follow-up clinic visit within 48 hours
- Consider referral to secondary care asthma clinic if 2nd attack within 12 months.

POOR RESPONSE
- Stay with patient until ambulance arrives
- Send written assessment and referral details
- Repeat β_2 bronchodilator via oxygen-driven nebulizer in ambulance

*** β_2 bronchodilator via spacer given one puff at a time, inhaled separately using tidal breathing; according to response, give another puff every 60 seconds up to a maximum of 10 puffs**

Figure 29.17 Management of acute severe asthma in children aged 2–5y^{G}

This figure is reproduced from *SIGN 158: British guideline on the management of asthma*, by kind permission of the Scottish Intercollegiate Guidelines Network.

⚠ *Lower threshold for admission if*
- Attack in late afternoon or at night
- Recent hospital admission or previous severe attack
- Concern over social circumstances or ability to cope at home

Acute abdominal pain

! Signs may be masked in elderly patients or those on corticosteroids. Small children with abdominal pain are difficult to assess.

History Consider:

- Site of pain—Figure 29.18
- Onset: how long? How did it start? Change over time?
- Character of pain: type of pain—burning, shooting, stabbing, dull, etc.
- Radiation
- Associated symptoms, e.g. nausea, vomiting, diarrhoea
- Timing/pattern, e.g. constant, colicky, relationship to food
- Exacerbating/relieving factors, including previous treatments and results
- Severity

Examination

- Temperature
- Pulse
- BP
- Jaundice
- Anaemia
- Site of pain (Figure 29.18)
- Guarding/rebound tenderness
- Rectal/vaginal examination as necessary

Management Treat the cause (Table 29.7)—if unsure admit as a surgical emergency to hospital. If possible, give analgesia prior to transfer to hospital.

Acute abdominal pain in pregnancy Non-obstetric causes of abdominal pain may be forgotten or signs may be less well localized than in the non-pregnant patient.

Appendicitis Mortality is higher in pregnancy and perforation more common (15–20%). Fetal mortality is 5–10% for simple appendicitis but rises to 30% when there is perforation. Due to the pregnancy, the appendix is displaced and pain is often felt in the paraumbilical region or subcostally. Admit immediately if suspected.

Cholecystitis Pregnancy encourages gallstone formation. Symptoms include RUQ pain, nausea, and vomiting. Diagnosis can be confirmed on USS. Treatment is the same as outside pregnancy aiming for interval cholecystectomy after birth.

Fibroids Torsion or red degeneration. Fibroids ↑ in size in pregnancy. They may twist if pedunculated. Red degeneration occurs usually after 20wk and may occur until the puerperium. It presents as abdominal pain ± localized tenderness ± vomiting and low-grade fever. Confirm diagnosis with USS. Treatment is with rest and analgesia. Pain resolves within 1wk.

Ovarian cysts Torsion or rupture of a cyst may both cause abdominal pain as may bleeding into a cyst. USS can confirm the presence of a cyst. Management depends on the nature of the cyst and the severity of the pain. Admit for assessment.

If >20wk gestation Also consider:

- Labour—➔ p. 812
- Pubic symphysis dehiscence—➔ p. 464
- Pre-eclampsia (epigastric pain)—➔ p. 804
- Haematoma of rectus abdominis
- Placental abruption—➔ p. 1084
- Uterine rupture—➔ p. 1084

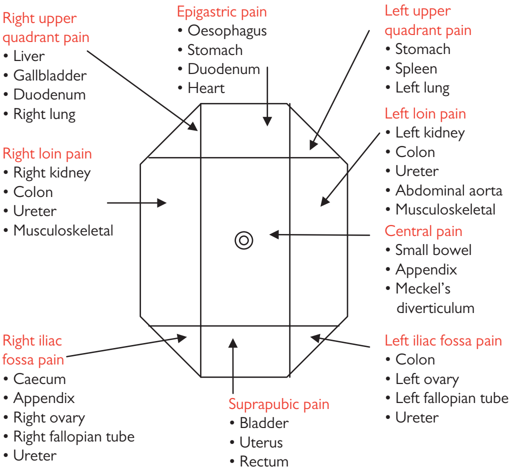

Right upper quadrant pain
• Liver
• Gallbladder
• Duodenum
• Right lung
Epigastric pain
• Oesophagus
• Stomach
• Duodenum
• Heart
Left upper quadrant pain
• Stomach
• Spleen
• Left lung
Right loin pain
• Right kidney
• Colon
• Ureter
• Musculoskeletal
Left loin pain
• Left kidney
• Colon
• Ureter
• Abdominal aorta
• Musculoskeletal
Central pain
• Small bowel
• Appendix
• Meckel's diverticulum
Right iliac fossa pain
• Caecum
• Appendix
• Right ovary
• Right fallopian tube
• Ureter
Suprapubic pain
• Bladder
• Uterus
• Rectum
Left iliac fossa pain
• Colon
• Left ovary
• Left fallopian tube
• Ureter

Figure 29.18 Site of abdominal pain gives important clues about the organ involved

Table 29.7 Differential diagnosis of acute abdominal pain

Renal and gynaecological causes			
Renal colic	➔ p. 418	Gynaecological malignancy	
UTI/pyelonephritis	➔ p. 422	Dysmenorrhoea	➔ p. 691
Urinary retention/hydronephrosis	➔ p. 428	Endometriosis	➔ p. 692
Henoch–Schönlein purpura	➔ p. 500	Pelvic inflammatory disease	➔ p. 714
Ovarian torsion/bleed/rupture	➔ p. 698	Ectopic pregnancy/ miscarriage	➔ p. 768, p. 767
GI causes			
Irritable bowel syndrome	➔ p. 388	Appendicitis	➔ p. 366
Constipation	➔ p. 350	Meckel's diverticulum	➔ p. 367
Diverticular disease	➔ p. 372	Pancreatitis	➔ p. 402
Gallbladder disease	➔ p. 400	Bowel obstruction/ischaemia	➔ p. 372
Inflammatory bowel disease	➔ p. 384	Intussusception	➔ p. 869
Gastroenteritis	➔ p. 348	Strangulated hernia	➔ p. 364
Gastritis/GORD	➔ p. 358	Volvulus	➔ p. 372
Peptic ulcer	➔ p. 360	GI malignancy	
Liver disease		Perforated bowel	
Other causes			
Muscular pain		Mesenteric adenitis	➔ p. 366
Spinal arthritis	➔ p. 450	Acute coronary syndrome	➔ p. 1066
Heart failure	➔ p. 232	Pneumonia	➔ p. 294
Sickle cell crisis	➔ p. 645	Subphrenic abscess	➔ p. 366
Ruptured spleen	➔ p. 1062	Diabetic ketoacidosis	➔ p. 1082
Torsion of the testis	➔ p. 440	Porphyria	➔ p. 599
Leaking/ruptured AAA	➔ p. 1060	Addison's disease	➔ p. 338
Shingles/post-herpetic neuralgia	➔ p. 628	Lead poisoning	

Fitting and funny turns

Transient loss of consciousness ➔ p. 524

Febrile convulsions ➔ p. 875

Epilepsy Adults—➔ p. 546; children—➔ p. 876

The fitting patient

When the call for assistance is received Instruct the attendant:

- To stay with the patient
- To turn the patient onto his/her side
- To move anything from the vicinity that might cause injury

If first fit Advise the attendant to call for an emergency ambulance immediately.

If known epilepsy Advise the attendant to call for an emergency ambulance if the fit does not stop spontaneously in <5min.

⚠ *Management of a major fit*

- Ensure that the airway is clear
- Turn the patient into the recovery position (Figure 29.4, ➔ p. 1043)
- Prevent onlookers from restraining the fitting patient
- Do not give drugs for the first 5min—many fits stop spontaneously

Medication For fits lasting ≥5min treat with:

- Midazolam buccal liquid:

>10y	10mg	*6mo–1y*	2.5mg
5–10y	7.5mg	*<6mo*	300 micrograms/kg
1–5y	5mg		

- *Or* rectal diazepam:

Elderly	10mg	*1mo–2y*	5mg
>10y	10–20mg	*Neonate*	1.25–2.5mg
2–10y	5–10mg		

- *Or* IV lorazepam (only use if resuscitation facilities are available):

Adult and child >12y	4mg	*Child <12y*	100 micrograms/kg—maximum 4mg

Status epilepticus Said to occur if a patient has more than one seizure without the patient regaining consciousness *or* fitting continues >5min despite medication.

Prolonged fitting If the fit continues >5min after medication administration, call for an emergency ambulance. Drugs can be repeated ×1 after 10–15min; consider checking finger prick blood glucose if prolonged fit.

Admit if

- Possibility that a fit is secondary to another illness, e.g. meningitis, subdural haematoma
- Recovery after the fit is incomplete (other than feeling sleepy)
- Status epilepticus (even if fits are controlled)

Follow-up Refer for urgent specialist assessment:

- Any adult who has a first fit
- Any child who has a first fit not related to fever

Acute stroke/TIA ➔ p. 534

Hypoglycaemia ➔ p. 1082

Delirium tremens (DTs) ➔ p. 1103

Vasovagal syncope (simple faint) ➔ p. 525

Acute hyperventilation/panic attack

Features Fear, terror, and feeling of impending doom accompanied by some or all of the following:

- Palpitations
- Shortness of breath
- Choking sensation
- Dizziness
- Paraesthesiae
- Chest pain/discomfort
- Sweating
- Carpopedal spasm

Differential diagnosis

- Dysrhythmia
- Asthma
- Anaphylaxis
- Thyrotoxicosis
- Temporal lobe epilepsy
- Hypoglycaemia
- Phaeochromocytoma (very rare)

Action

- Talking down Explain the nature of the symptoms to the patient:
  - Racing of the heart is due to adrenaline produced by the panic
  - Paraesthesiae/feelings of dizziness are secondary to overbreathing due to panic
  - Count breaths in and out gently slowing breathing rate
- Rebreathing techniques:
  - Place a paper bag over the patient's mouth, and ask him/her to breathe in and out through the mouth
  - A connected but not switched on O_2 mask or nebulizer mask is an alternative in the surgery
  - This raises the partial pressure of CO_2 in the blood, symptoms due to low CO_2 (e.g. tetany, paraesthesiae, dizziness) resolve, and also demonstrates the link between hyperventilation and symptoms
- Propranolol 10–20mg stat may be helpful—DO NOT USE for patients with asthma or heart failure, or those taking verapamil

Recurrent panic attacks ➔ p. 972

Further information

NICE (2010, updated 2014) Transient loss of consciousness ('blackouts') in over 16s. 🖱 www.nice.org.uk/guidance/cg109

NICE (2012, updated 2018) The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care. 🖱 www.nice.org.uk/guidance/cg137

Endocrine emergencies

Hypoglycaemia Usually known to have diabetes and taking insulin, an injectable GLP-1 mimetic (e.g. liraglutide), or oral medication (e.g. sulfonylurea, gliptin, or rapid acting secretagogue). Short history. May present with coma, fits or odd/violent behaviour, tachycardia ± ↑ BP. There may have been warning signs/symptoms—sweating, hunger, tremor. ❗ Younger children may present atypically with behavioural changes or headache.

Investigation Capillary blood glucose <4mmol/L (usually <2.5mmol/L).

⚠ *Action* Is the patient able to self-treat?

- **If yes** Give 10–20g of rapid-acting carbohydrate, e.g. 3 glucose tablets, oral glucose gel (e.g. Glucogel®), 2–3 teaspoons of sugar, 100mL of milk, a sugar-containing soft drink (e.g. Lucozade®), or 5 sweets (e.g. Jelly Babies®)
- **If no** Give IM glucagon 1mg (children <25kg—0.5mg). ❗ May have poor effect if starved or drunk. Alternatively (if IV access is possible and equipment available), give IV glucose (*adult*: 50–250mL of 10% solution in 50mL aliquots; *child*: 2–5mL/kg of 10% solution)
- **Once the patient has regained consciousness** Give simple carbohydrate as for the conscious patient and, as symptoms improve, give complex carbohydrate, e.g. 2–3 biscuits or 2 slices of bread
- **Repeat glucose testing** In 15min then monitor hourly over the next 4h and then every 4h for the following 24h. Maintain a high glucose intake for several hours if hypoglycaemia due to a sulfonylurea or gliptin
- **Review reasons for the hypoglycaemia** (➔ p. 324)
- **Admit** To hospital if capillary blood glucose remains <4mmol/L despite treatment in the community; the patient does not regain consciousness even though capillary blood glucose is >4mmol/L; or the patient is unable to self-monitor at home or has poor social support

Hyperglycaemic ketoacidotic coma Usually patients with type 1 DM but may occur in patients with type 2 DM, especially those being treated with GLP-1 mimetics (e.g. exenatide, liraglutide) or gliflozins (e.g. canagliflozin). May be the way in which type 1 DM presents.

Presentation 2–3d history of deterioration, often precipitated by infection. Typically the patient is dehydrated with Kussmaul breathing (deep sighing breaths), ketotic (fruity) smelling breath, shock (↓ BP and postural drop, tachycardia) ± coma. ⚠ Can present with vomiting and abdominal pain—always check for ketotic breath and Kussmaul breathing.

Investigation Capillary blood glucose is usually >20mmol/L and urine (if available) tests +ve for ketones. ❗ Gliflozins (e.g. canagliflozin, dapagliflozin, empagliflozin) may cause diabetic ketoacidosis even with normal blood glucose—warn patients.

⚠ *Action* Arrange to admit as an emergency to hospital. If shocked/coma—lie flat, elevate feet, and resuscitate:

- Airway—check airway is clear
- Breathing—give O_2 if available to maintain SpO_2 at 94–98%
- Circulation—gain IV access if possible and give 1L (child: 10mL/kg) 0.9% saline rapidly. Repeat up to 3× as needed

Hyperosmolar hyperglycaemic state Also known as hyperglycaemic hyperosmolar non-ketotic coma (HONK). Predominantly occurs in patients with type 2 DM (very rarely type 1). Presents with <1wk history of deterioration, ↓ level of consciousness, dehydration ++, ↓ BP with postural drop. Often precipitated by other illness, e.g. infection, MI. May be a presenting feature of type 2 DM. Blood sugar (on blood testing strip) is >35mmol/L.

⚠ *Action* Admit to hospital as an acute medical emergency.

Myxoedema (hypothyroid) coma Rare. Typically occurs in patients >65y and may be precipitated by intercurrent illness (e.g. MI, stroke, infection) or trauma. The patient may look hypothyroid, or have a history of thyroid surgery or treatment with radioactive iodine.

Presentation Hypothermia, heart failure, hyporeflexia, hypoglycaemia, bradycardia, cyanosis, seizures, coma.

⚠ *Action* If suspected, admit as an acute medical emergency. While awaiting admission, keep warm, give O_2 to maintain peripheral oxygen saturations between 94–98% and, if needed, treat heart failure with IV furosemide (➔ p. 1068).

Hyperthyroid crisis (thyrotoxic storm) Rare. Risk factors include recent thyroid surgery/history of radioactive iodine and/or intercurrent illness (e.g. MI, infection) or trauma.

Presentation Fever, agitation, confusion, diarrhoea/vomiting, acute abdomen, tachycardia ± AF, may have goitre ± thyroid bruit, coma.

⚠ *Action* Admit to hospital as an acute medical emergency.

Hypoadrenal (Addisonian) crisis

Presents with vomiting, hypotension, and shock. May occur:

- In patients on long-term steroids (treatment or replacement) if the steroids are stopped suddenly or not ↑ during intercurrent illness, *or*
- As a presenting feature of congenital adrenal hyperplasia or Addison's disease

Management Admit to hospital as an acute medical emergency. If suspected, give IM or IV hydrocortisone while awaiting admission:

- Adults and children >12y 100mg
- Children 1mo–12y 2–4mg/kg

Prevention of Addisonian crises

- Warn all patients taking long-term steroids not to stop their steroids abruptly and to tell any doctor treating them about their condition
- Advise patients to carry a steroid card or Addison's disease self-help group emergency card, and wear a MedicAlert® bracelet or similar in case of emergency
- Double dose of steroid prior to dental treatment or if intercurrent illness (e.g. URTI)
- If vomiting, replace oral steroid with IM hydrocortisone

Obstetric emergencies

Resuscitation of the newborn ➔ p. 1086

Eclampsia Occurs when a pregnant woman has a fit as a result of pre-eclamptic toxaemia (PET)—➔ p. 804. Usually BP is very high and, if the baby is not yet born, it becomes distressed. There is a serious risk of stroke in the mother. Women with pre-eclampsia have a 2% chance of eclamptic seizure. 44% occur after the baby is born—usually <24h after delivery. Give buccal midazolam (10mg) or PR diazepam (10–20mg) or IV lorazepam (4mg) and admit as an acute 'blue light' emergency.

HELLP syndrome Occurs in pregnancy or <48h after delivery: Haemolysis, Elevated Liver enzymes, and Low Platelets. Associated with severe pre-eclampsia. Presents with hypertension (80%); right upper quadrant pain (90%); nausea and vomiting (50%) ± oedema. Admit as an emergency for obstetric assessment.

Obstetric shock *Causes*: haemorrhage—APH (➔ p. 810); placental abruption (remember—bleeding may be internal and not seen per vaginum); PPH (➔ p. 811); ruptured or inverted uterus; amniotic fluid embolism; broad ligament haematoma; pulmonary embolus; anaphylaxis; septicaemia.

> ⚠ *Action* Arrange immediate emergency admission to the nearest specialist obstetric unit or A&E. If possible, gain IV access and start IV fluids; give O_2 via face mask. Treat the cause if apparent.

Amniotic fluid embolism Rare. Mortality ~80%. Presents with shock, cyanosis, and dyspnoea. May occur at the height of a contraction. If suspected call for help; resuscitate—Airway; Breathing; Circulation; and transfer as an emergency to the nearest A&E or obstetric unit.

Fetal distress Signifies hypoxia. Suggested by passage of meconium during labour, and fetal tachycardia (>160bpm at term) or bradycardia (<100bpm). Give the mother O_2 via face mask and turn her onto her side. Transfer immediately by ambulance to a specialist obstetric unit.

Acute abdominal pain in pregnancy ➔ p. 1078

Uterine rupture Rare in the UK. Associated with maternal mortality of 5% and fetal mortality of 30%. 70% are due to dehiscence of Caesarean section scars. Rupture occurs most commonly during labour but occasionally in the 3rd trimester. Usually presents with severe, bursting, constant lower abdominal pain ± heavy vaginal bleeding. Associated with profound shock in the mother and fetal distress. If in labour, the presenting part may disappear from the pelvis ± contractions stop. If suspected, admit as an acute emergency to a specialist obstetric unit

Placental abruption (abruptio placentae) Part of the placenta becomes detached from the uterus. Consequences depend on the degree of separation and the amount of blood loss. Typically presents with constant pain—may be felt in the back if posterior placenta. The uterus is tender and feels woody hard. Often accompanied by shock ± PV bleeding, and absent fetal heart or signs of fetal distress (fetal tachycardia/bradycardia). If suspected, admit as an acute emergency to a specialist obstetric unit.

Shoulder dystocia Affects <1% deliveries but is a life-threatening emergency. Occurs when the anterior shoulder impacts upon the symphysis pubis after the head has delivered and prevents the rest of the baby following. Most cases of shoulder dystocia are unanticipated. *Clues*: prolonged 1st or 2nd stage of labour and 'head bobbing'—the head consistently descends then returns to its original position during a contraction or pushing in the 2nd stage.

⚠ *Action* ❗ If shoulder dystocia occurs in the community there is usually no time to transfer a woman to a specialist unit. Call for emergency ambulance support. Consider episiotomy. Then try (in no particular order):

- Roll the mother onto hands/knees and try delivering the posterior shoulder
- Flex and abduct the mother's legs up to her abdomen (upside down squatting position)—try delivery again
- Deliver the posterior arm—put a hand in the vagina in front of the baby—ensure the posterior elbow is flexed in front of the body and pull to deliver the forearm. The anterior shoulder usually follows
- External pressure—ask an assistant to apply suprapubic pressure with the heel of the hand—a rocking movement can help
- Adduction of the most accessible (preferably anterior) shoulder. Simultaneously put pressure on the posterior clavicle to turn the baby. If unsuccessful, continue rotation through 180 degrees and try again

Cord prolapse The cord passes through the os in front of the presenting part of the baby. If the presenting part squashes the cord, umbilical blood flow is restricted causing fetal hypoxia and distress (fetal mortality 10–17%). Risk factors include: malpresentation; cephalo-pelvic disproportion; multiple pregnancy; preterm rupture of membranes; polyhydramnios; pelvic tumours.

⚠ *Action* Admit as an emergency to the nearest specialist obstetric unit. While awaiting hospital transfer, try to keep the cord in the vagina and minimize handling of the cord to ↓ cord spasm. Try to prevent the presenting part from occluding the cord by displacing the presenting part upwards with the examining hand, turning the patient into the knee/elbow position with head down and/or dropping the head of the bed.

Retained placenta The 3rd stage of labour is complete in <10min in 97% of labours. If the placenta has not been delivered in <30min (to allow for cervical spasm), it is likely not to deliver spontaneously. Avoid excessive cord traction. Check the placenta is not in the vagina—remove if it is. Check the uterus. If the uterus is well contracted, cervical spasm is probably trapping an otherwise separated placenta—wait for cervix to relax to enable removal of the placenta. If the uterus is bulky, the placenta may have failed to separate. *Try*: rubbing up a contraction; putting the baby to the breast (stimulates uterine contraction); and/or giving a further dose of syntometrine. If the placenta will still not deliver, admit as emergency for manual removal.

Uterine inversion Rare. Do not try to remove the placenta if still attached. Admit as an emergency. The mother may become profoundly shocked so set up an IV infusion before transfer if possible, and give O_2 via a face mask.

Broad ligament haematoma Presents in a recently delivered woman as obstetric shock without excessive PV bleeding. Examination reveals pain and tenderness on the affected side. The uterus is deviated from that side. Admit as an acute emergency to the nearest specialist obstetric unit.

Resuscitation of the newborn

Follow the algorithm in Figure 29.19.

Rapid assessment of the infant at birth

Start the clock. Assess colour, tone, breathing, heart rate.

Healthy baby

- Born blue
- Good tone
- Good heart rate (120–150bpm)
- Rapidly becomes pink during the first 90s
- Cries seconds after delivery

Ill baby

- Born pale
- Poor tone/floppy
- Slow/very slow heart rate (<100bpm)
- Not breathing/inadequate breathing by 90–120s

Keep the baby warm Newborn babies develop hypothermia very quickly and this can affect outcome. Dry quickly and cover.

Clamping the cord ≥1min after delivery for newborns not requiring resuscitation. If resuscitation is needed, delay until the baby is breathing.

Heart rate and pulse oximetry Heart rate is best judged by listening with a stethoscope—in many cases it can also be felt by palpating the umbilical cord. If available and the infant needs resuscitation, use pulse oximetry to measure pre-ductal oxygen saturation and heart rate. Place sensor on the right hand/wrist. In healthy term babies, oxygen saturation ↑ from 60% soon after birth to 90% at 10min.

Airway Open the airway by placing the head in a neutral position (neck neither extended nor flexed). If the occiput is prominent and the neck tends to flex, place a support under the shoulders—but do not overextend the neck. If the baby is floppy, apply jaw thrust or chin lift as needed.

Breathing Inflation breaths are breaths with pressures of ~30cm of water for 2–3s. Use air rather than oxygen.

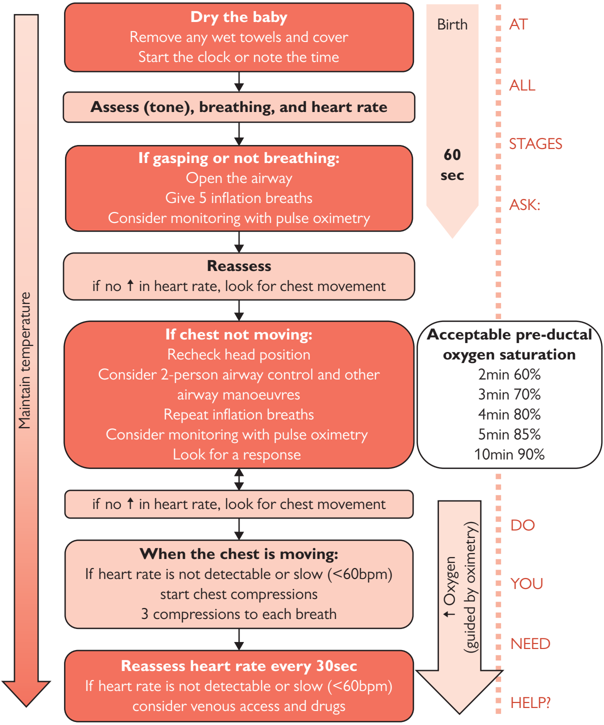
- If heart rate ↑ You have successfully inflated the chest. If the baby does not start breathing alone, continue to provide regular breaths at a rate of ~30–40 breaths/min until the baby starts to breathe alone
- If heart rate does not ↑ Either you have not inflated the chest or the baby needs more help. If the chest does not move, consider:
  - Is the baby's head in the neutral position? Do you need jaw thrust?
  - Do you need a longer inflation time?
  - Do you need a second person's help with the airway?
  - Is there obstruction, e.g. meconium (laryngoscope and suction)?
  - What about an oropharyngeal (Guedel) airway?

Chest compressions Only commence after inflation of the lungs.

- Grip the chest in both hands so that the thumbs of both hands can press on the sternum at a point just below an imaginary line joining the nipples and with the fingers over the spine at the back
- Compress the chest quickly—↓ the AP diameter of the chest by at least a third with each compression. Ratio of compressions to inflations is 3:1

Drug support For a few babies inflation of the lungs and effective chest compression are not sufficient to produce effective circulation. IV or intraosseous drugs may be helpful. Consider:

- Adrenaline (epinephrine) 10 microgram/kg (0.1mL/kg of 1:10,000 solution), increasing to 30 micrograms/kg (0.3mL/kg of 1:10,000 solution) if ineffective
- Glucose 250mg/kg (2.5mL/kg of 10% glucose)
- For emergency volume replacement (e.g. history of a bleed)—use 10mL/kg 0.9% saline given over 10–20s. Repeat if needed



Dry the baby
Remove any wet towels and cover
Start the clock or note the time

Assess (tone), breathing, and heart rate

If gasping or not breathing:
Open the airway
Give 5 inflation breaths
Consider monitoring with pulse oximetry

Reassess
if no ↑ in heart rate, look for chest movement

If chest not moving:
Recheck head position
Consider 2-person airway control and other airway manoeuvres
Repeat inflation breaths
Consider monitoring with pulse oximetry
Look for a response

if no ↑ in heart rate, look for chest movement

When the chest is moving:
If heart rate is not detectable or slow (<60bpm) start chest compressions
3 compressions to each breath

Reassess heart rate every 30sec
If heart rate is not detectable or slow (<60bpm) consider venous access and drugs

Acceptable pre-ductal oxygen saturation
2min 60%
3min 70%
4min 80%
5min 85%
10min 90%

Figure 29.19 Newborn life support algorithm

Reproduced with the kind permission of the Resuscitation Council (UK).

Further information

Resuscitation Council (UK) (2015) Resuscitation and support of transition of babies at birth. www.resus.org.uk/resuscitation-guidelines/resuscitation-and-support-of-transition-of-babies-at-birth

Accidents and injuries

Road accidents Doctors are not legally obliged to attend an accident they happen to pass—but most feel morally obliged to do so.

Immediate action

- Assess the scene
- Ensure that emergency services have been called
- Take steps to ensure your own safety and that of others, e.g. park your vehicle defensively; wear a reflective jacket if available; turn on hazard lights; use warning triangles
- Ensure all vehicle ignitions are turned off and forbid smoking
- Triage casualties into priority groups—decide who to attend first

First aid treatment

- Check the need for basic resuscitation: Airway patent? Breathing adequate? Circulation intact? Resuscitate as necessary (Adult—➔ p. 1040; Child—➔ p. 1044)
- Control any haemorrhage with elevation and pressure
- DO NOT attempt to move anyone who potentially could have a back or neck injury until skilled personnel and equipment are available
- Do not give anything by mouth
- Use coats and rugs to keep victims warm
- If available give analgesia (e.g. opioids—but not if significant head injury or risk of intraperitoneal injury; Entonox®—from ambulance)
- If shocked, set up IV fluids if available
- Take directions from the paramedics—they are almost certainly more experienced than you in these situations

Medicolegal issues

- Ensure your medico-legal insurance covers emergency treatments
- Keep full records of events, action taken, drugs administered, origin of drugs, batch numbers, and expiry dates
- A GP can charge a fee to the victims for any assistance given

Road safety 30–40% of all fatal accidents are road accidents. *Prevention*:

- Supervise children close to roads; teach them the Green Cross code
- Wear seat belts and appropriate protective clothing (e.g. helmet if riding a pedal or motorcycle)
- Ensure that children have car seats appropriate for their age/height and are properly strapped in
- Avoid alcohol/other drugs that hamper performance when driving
- Do not drive if tired or ill
- If cycling, use cycle tracks if available
- Keep vehicles well maintained
- Keep speed down

Home safety Every year >4000 people die due to accidents in the home and nearly 3 million seek treatment in A&E departments. Spot the dangers; offer safety advice (e.g. from health visitor if young children in the house); advise patients to fit smoke alarms and safety devices (e.g. stair gates for toddlers); ensure adequate supervision of children or elderly confused people; maintain equipment correctly.

Head and facial injury ➔ p. 1094

Routine vaccination schedule

Primary immunization 3 doses with 1mo between each (i.e. 2nd dose one month after 1st dose and 3rd dose one month after 2nd dose

Booster doses 1 booster dose 5y after primary immunization (3y if child aged <10y) and a second booster dose 10y after the first booster dose

Tetanus-prone injuries

- Puncture-type injuries acquired in a contaminated environment and likely to contain tetanus spores, e.g. gardening injuries
- Wounds containing foreign bodies, e.g. splinters
- Compound fractures
- Wounds/burns with systemic sepsis
- Certain animal bites and scratches from animals that have been rooting in soil, or agricultural animals

High risk Heavy contamination with material likely to contain tetanus spores; extensive devitalized tissue and/or wounds awaiting surgical intervention delayed for >6h

Immunization status	**Immediate treatment**		
	Clean wound	***Tetanus-prone wound***	***High risk wound***
• Aged ≥11y—received primary immunization and last booster dose <10y ago • Aged 5–10y—received primary immunization and pre-school booster • Aged <5y—received primary immunization	None	None	None
• Aged ≥11y—received primary immunization but last booster dose ≥10y ago • Aged 5–10y—received primary immunization but no pre-school booster	None	Give vaccine	Give vaccine + Tetanus Ig (TIG)*
• Not received primary immunization • Uncertain immunization status (including if born before 1961)	Give vaccine	Give vaccine + TIG*	Give vaccine + TIG*

⚠Give further doses of vaccine as required to complete the recommended schedule (to ensure future immunity)

* Human normal immunoglobulin (HNIG) is an alternative.

Figure 29.20 Who should have tetanus vaccination?[G]

Burns and scalds ➔ p. 1096
Poisoning and overdose ➔ p. 1098
Muscle injuries/sprains ➔ p. 476

Drowning ➔ p. 1108
Fractures ➔ p. 1092
Whiplash ➔ p. 449

Haematoma Subungual—➔ p. 461; pinna—➔ p. 1095

Wounds Most patients with significant lacerations present directly to A&E. If a patient presents to general practice, perform immediate care (elevate bleeding limb and apply pressure). Advise nil by mouth and transfer to A&E.

Minor lacerations

- Check tetanus status (Figure 29.20)
- Ensure no foreign body is in the wound—if in doubt refer for X-ray/surgical exploration (especially important if the injury was with glass)
- Wash the wound and clean away debris and any necrotic material
- Check there is no damage to underlying nerves, tendons, bone, or blood supply before dressing or closing the wound

- Aim to oppose the skin edges without tension to allow healing
- Do not attempt to close a wound if you are not confident that you can achieve an adequate result
- Always refer cuts through the lip margin to A&E; consider referral to A&E for any facial wounds and wounds in children
- In assault cases take particular care to document all injuries carefully, e.g. with photographs, drawings, and measurements of wounds
- Consider non-accidental injury in children—➲ p. 902

Closing the wound Options:

- **Skin closure strips** Use for small cuts in non-hairy skin not under tension or in addition to sutures for larger wounds
- **Skin 'glue' (e.g. Histoacryl®)** Quick (takes 30s to set) and can be used on hairy skin such as the scalp
- **Suturing** Undertake training before attempting suturing:
  - Infiltrate wound edges with 1% lidocaine (maximum 2mg/kg)
  - Addition of adrenaline (epinephrine) can help haemostasis but must not be used on digits or extremities, as necrosis can occur
  - Take care to oppose edges accurately—start interrupted sutures in the middle of the wound
  - Use appropriate suture (e.g. monofilament nylon: adult face 5–0—remove after 5d; limbs or trunk 3–0—remove after 1–2wk)

Pretibial lacerations The shin has poor blood supply especially in the elderly. Flap wounds are common, may heal poorly ± break down to form ulcers.

Management Wash the wound. Carefully realign the flap; secure with skin closure strips without tension, and bandage. Advise elevation of the leg. Review regularly to check healing.

Airgun pellets Common. Refer for X-ray. Can be difficult to remove—leave in place if not in a harmful position. If in a joint, refer for removal.

Fish hooks Infiltrate with lidocaine. Push the hook forwards through the skin until the barb is exposed. Cut the barb off and then ease the hook back through the skin the same way it entered.

Knocked-out teeth Ask the patient to suck the tooth clean, reinsert, or store in milk or saliva and send to a dentist.

Coin and other foreign body ingestion Most coins will pass through the gut without any problems. If asymptomatic, they can be left to take their course (advise checking stools to ensure passed). If symptomatic, refer for X-ray and consideration for endoscopic removal. If there is any indication of aspiration refer urgently.

Foreign bodies in the ear Most common in children. If the patient is cooperative, try to remove under direct vision with forceps but avoid pushing objects deeper into the canal and causing damage; removal under GA may be needed. Insects can be drowned in oil and syringed out.

Foreign bodies in the nose Common in young children. Any child with smelly discharge from one nostril has a foreign body in the nose until proven otherwise—refer for exploration under GA. Do not try to remove yourself unless the object is very superficial and the child cooperative. You might push the object further in and cause trauma.

Removal of ticks Use a commercially available tick remover when possible. If a tick remover is not available, grip the tick as close as possible to the skin with a pair of tweezers, and firmly pull the tick out of the skin.

Animal bites ~200,000 people are bitten by dogs each year in the UK. Animal bites are contaminated and wound infection is common. Clean with soap and water. Check tetanus status. Do not suture unless cosmetically essential and there is minimal tissue damage—refer if in doubt. Give prophylaxis against infection (e.g. co-amoxiclav or erythromycin + metronidazole).

Human bites These are especially prone to infection. Also consider risk of hepatitis B and HIV. If HIV prophylaxis is indicated, it needs to be started immediately—refer urgently to A&E for local policy implementation.

Snake bites The adder is the only poisonous snake in the UK. Bites are only rarely lethal. Attempt to identify the snake and refer the patient urgently to hospital. Do not apply a tourniquet or cut/suck the wound.

Rabies risk^{G} Consider post-exposure prophylaxis for rabies (vaccination ± immunoglobulin) if:

- Any exposure to bat secretions or bat bites in the UK or elsewhere (people who work with bats should be vaccinated prior to exposure)
- Animal bite or exposure to animal secretions elsewhere in the world—risk depends on the location

Insect stings Reactions range from blisters through papules to urticarial wheals—2° infection is common.

- **Anaphylaxis** Follow algorithm in Figure 29.10, ➔ p. 1055 and admit to hospital as a blue light emergency
- **Immediately after the sting** Remove any sting present in the wound; often no further treatment is needed
- **If severe local reaction occurs** Apply an ice pack; give oral antihistamine (e.g. chlorphenamine 4mg stat); continue antihistamine 4–6-hourly as needed
- **If 2° bacterial infection** Treat with oral or topical antibiotics
- **Remove sources of insects** e.g. remove fleas from carpets with household flea spray (multiple bites on ankles and lower legs)

Weaver fish sting Common on sandy beaches. The fish lurks under the sand and so is usually trodden on—presents with severe pain in the foot. Immerse the affected area in uncomfortably hot (but not scalding) water. Give analgesia. Pain resolves after 2–3d.

Jellyfish sting Remove the patient from the sea as soon as possible. Scrape or wash adherent tentacles off. Alcoholic solutions including suntan lotions should not be applied because they may cause further discharge of stinging hairs. Ice packs ↓ pain and a slurry of baking soda (sodium bicarbonate), but not vinegar, may be useful for treating stings from UK species.

Further information

Public Health England (2018) The Green Book: rabies. 🖱 www.gov.uk/government/publications/rabies-the-green-book-chapter-27

Public Health England (2018) The Green Book: tetanus. 🖱 www.gov.uk/government/publications/tetanus-the-green-book-chapter-30

Fractures

Bony fractures usually present with pain in the affected bone following injury and ↓ function. On examination there may be bruising/swelling, deformity, local tenderness, impaired function ± crepitus/abnormal mobility.

⚠ Action if suspected fracture

- Immobilize the affected bone and give analgesia
- Refer to A&E for X-ray and further management
- If available and the patient is shocked, start an IV infusion of 0.9% saline while awaiting hospital transfer

Ottawa rules for ankle or foot injury Foot and ankle injuries are common. It can be difficult to distinguish between a sprain and a fracture. The Ottawa rules ↓ need for X-ray by ~25%:

Ankle injury Refer for an ankle X-ray if there is pain in the malleolar area AND:

- Bone tenderness at the posterior tip of the lateral malleolus, *or*
- Bone tenderness at the posterior tip of the medial malleolus, *or*
- Unable to weight bear at the time of the injury and when seen

Foot injury Refer for a foot X-ray if there is pain in the mid-foot AND:

- Bone tenderness at the 5th metatarsal base, *or*
- Bone tenderness at the navicular, *or*
- Unable to weight bear at the time of injury and when seen

Otherwise diagnose a sprain Treat sprains with rest, ice, compression, elevation, and analgesia (paracetamol ± NSAIDs). If severe (or the patient is an athlete), refer to physiotherapy.

Fractures See Table 29.8.

Always consider assessment and treatment for osteoporosis in all men and women >50y who have had a fragility fracture (Colles' fracture, hip fracture, and/or vertebral collapse).

Head and facial injury ➔ p. 1094

Fracture complications Often occur after the patient has been discharged from hospital and may present as a primary care emergency. *Refer back to the fracture clinic or A&E if*:

- Persistent pain
- Offensive odour or discharge
- Limb swelling that is not settling
- External pins/wires become infected
- Wound infection that is not settling with oral antibiotics
- External pins/wires are catching on clothing or other parts of the body
- Cast edges are abrading the skin
- Cast has deteriorated in structural strength, e.g. from getting wet

Compartment syndrome Crush injury, fracture, prolonged immobility, or tight splints, dressings, or casts can result in ↑ pressure within muscle compartments and eventually vascular occlusion. Presents with swelling, severe pain (↑ on passive stretch of muscles), distal numbness, redness, mottling, blisters. ❗ Pulses may be present distally. Loosen any restricting bandage/cast. Refer as an emergency to orthopaedics—fasciotomy may be needed to relieve the pressure.

Table 29.8 Common fractures seen in primary care

Fracture	Features and management
Clavicle	Common injury (5% all fractures). Usually results from a fall onto an outstretched arm. 80% fractures are in the middle third; 15% the lateral third; and 5% the medial third ***Management*** Refer to A&E for confirmation of diagnosis and fracture clinic follow-up. Treatment is with sling support and analgesia ***Complications*** Pneumothorax, malunion, and nerve/vessel damage
Colles'	Most commonly due to a fall onto an outstretched hand in an elderly ♀. Presents with pain and swelling of the wrist ('dinner fork' deformity) ***Management*** Refer any suspected fracture for X-ray and reduction ***Complications*** Include rupture of the extensor pollicis longus tendon, carpal tunnel syndrome, and reflex sympathetic dystrophy
Scaphoid	Caused by falling onto an outstretched hand. Presents with pain, swelling, and tenderness in the anatomical snuffbox. Symptoms may be mild and fracture is easily missed Refer suspected cases for scaphoid view X-rays. If X-ray is inconclusive and pain continues, repeat 2wk later—bone scan or MRI can help if still –ve ***Complications*** Non-union and avascular necrosis of the proximal fragment, which can lead to long-term problems of arthritis and pain
Fingers	Common injuries. Often associated with sport. ***Management*** Refer all suspected fractures for X-ray ± reduction
Hip	Common among the elderly and carries high morbidity and mortality (~25%). ♀ > ♂. Usually occurs through the neck of the femur. Risk factors include: maternal hip fracture, osteoporosis, unsteadiness, sedative medication, poor eyesight, and polypharmacy There may be a history of a fall but not always. Suspect in any patient who is elderly or has risk factors for osteoporosis who is 'off legs'. Occasionally, patients can still weight bear with difficulty. Signs include: external rotation, shortening, and adduction of leg ***Management*** Refer urgently to A&E for X-ray
Ankle	History is of a fall over an obstacle or trip down a step. The ankle rapidly becomes swollen and tender—often bilaterally ***Management*** Decide whether an X-ray is needed (see Ottawa rules, ➔ p. 1092). If so, refer to A&E
Metatarsals	The most common fracture is of the base of the 5th metatarsal in an 'ankle twisting' injury. March or stress fractures occur in people who do a lot of walking or running and affect the neck/shaft of the 2nd metatarsal ***Management*** Decide whether an X-ray is needed. If so, refer to A&E. Undisplaced fractures are usually treated with analgesia and support
Toes	Caused by stubbing the toe or dropping a heavy object on it. ***Management*** • *Undisplaced suspected fracture* Do not X-ray unless diagnosis is in doubt. Support the injured toe by 'buddy' strapping it to the adjacent toe. Give analgesia • *Displaced fracture and/or dislocation* Refer for X-ray and reduction

Head and facial injury

⚠ Severe head injury

- Perform basic life support (adult ➔ p. 1040; child ➔ p. 1044)
- Protect the cervical spine
- Transfer to A&E by ambulance

Less severe head injuries

History If possible take the history from a witness as well as the patient. Ask about circumstances of injury, loss of consciousness (LOC), seizures, current symptoms, and behaviour.

Examination Check scalp and head for injury, perform a brief neurological examination (including fundi), and check for other injuries—accompanying neck injuries are common.

⚠ Refer to A&E if[N]

- Glasgow Coma Scale <15 at any time since injury—➔ p. 1052
- Any LOC since injury
- Focal neurological deficit since injury—problems speaking, understanding, reading, or writing; ↓ sensation; loss of balance; weakness; visual changes; abnormal reflexes; problems walking; irritability; or altered behaviour—especially in young children
- Any suspicion of skull fracture; penetrating head injury; blood or CSF in the nose, ear, or wound; serious scalp laceration or haematoma
- Amnesia for events before or after injury
- Vomiting since injury
- Persistent headache since injury
- Seizure since injury
- Any previous cranial neurosurgical interventions
- High-energy head injury (e.g. pedestrian hit by motor vehicle, fall >1m or fall >5 stairs)
- History of bleeding or clotting disorder or on anticoagulant therapy
- Difficulty in assessing the patient (e.g. very young, elderly, intoxicated, or epileptic) or concern about diagnosis
- Suspicion of non-accidental injury
- Inadequate supervision at home

❗ If neck pain/tenderness, focal neurological deficit, paraesthesiae in the extremities, or any other clinical suspicion of cervical spine injury, immobilize the neck and refer to A&E.

If examination is normal

- Warn the patient (+ carer) that he/she may suffer mild headaches, tiredness, dizziness, tinnitus, poor concentration, and poor memory for the next few days
- Advise rest and paracetamol (but not codeine) for the headache
- Young children can be difficult to assess—sleepiness is common and not a worrying sign as long as the child is rousable
- Give written head injury information regarding warning signs to trigger reconsultation—drowsiness, severe headache, persistent vomiting, visual disturbance, and/or unusual behaviour

Cervical spine injury If spinal cord injury is suspected (e.g. if the victim has sustained a fall, been struck on the head or neck, or has been rescued after diving into shallow water) take particular care during handling and resuscitation to maintain alignment of the head, neck, and chest in the neutral position. A spinal board and/or cervical collar should be used if available.

Whiplash injury ➔ p. 449

Injury to the face Mostly due to road accidents or violent incidents. Carefully document injuries as your notes may be required for legal proceedings. Look for other injuries, e.g. airway problems, head injury, neck injury. Palpate the face for signs of a fracture—if present refer to maxillofacial surgeons for assessment. Check tetanus status. Post-traumatic stress (➔ p. 976) is common after facial injury.

Specific injuries

- **Facial lacerations** Refer to A&E for suturing
- **Fractured mandible** A blow to the jaw can cause unilateral or bilateral fractures. Presents with pain (worse on moving jaw), bruising ± bleeding inside the mouth ± discontinuity of the teeth (displaced fracture) ± numbness of the lower lip (if the inferior dental nerve has been damaged). Refer to A&E for X-ray and further management
- **Dislocated jaw** Presents with pain and the mouth is stuck open—refer to A&E or maxillofacial surgeons for X-ray and reduction
- **Fractured zygoma/malar complex** A blow on the cheek may fracture the zygomatic arch in isolation or more usually cause a 'tripod' fracture. *Signs*: bony tenderness, flattening of the malar process—best seen from above (may be masked by swelling), epistaxis, subconjunctival haemorrhage extending posteriorly, and infraorbital numbness ± jaw locked. Refer to A&E for X-ray and further management. Advise not to blow nose
- **Middle third facial fractures (Le Fort)** Usually bilateral. *Signs*: epistaxis, CSF rhinorrhoea, crepitus on palpation, swelling, open bite, and risk of airway compromise. Refer to A&E for X-ray and further management
- **Haematoma of the pinna** Usually follows trauma (e.g. rugby injury). Must be evacuated urgently (aspirated via large-bore needle or surgically) to prevent necrosis of the cartilage and 'cauliflower' ear—refer to A&E or ENT
- **Nasal fracture and other nasal injuries** ➔ p. 919
- **'Blow out' fracture of orbit** ➔ p. 937
- **Avulsed tooth** ➔ p. 1090
- **Dog bite** ➔ p. 1091

Post-concussion syndrome Seen following even quite minor head injury. Due to neuronal damage. Features include all, or some, of:

- Headache
- Dizziness
- Fatigue
- Depression
- Poor concentration
- Memory problems

Treatment is supportive and symptoms usually resolve with time (but can take months or even years).

Further information

NICE (2014, updated 2017) Head injury: assessment and early management. 🖰 www.nice.org.uk/guidance/cg176

Scalds and burns

Assess

- Cause, size, and thickness of the burn
- Use the 'rule of nines' to estimate the extent of the burn (Box 29.4)
- Partial-thickness burns are red, painful, and blistered; full-thickness burns are painless and white or grey
- Always consider non-accidental injury in children—➔ p. 902

⚠ Immediate action

- Remove clothing from the affected area and place under cold running water for >10min or until pain is relieved
- Do not burst blisters
- Prescribe/give analgesia

Refer All but the smallest partial thickness burns for assessment in A&E (*adult*: ≥3% total body surface area; *child*: ≥2% total body surface area). If referring to A&E, consider covering burns with cling film prior to transfer (transparent, sterile, and non-stick) In particular, refer all:

- Burns in special areas—face, hand, perineum/genitals, feet, flexures (particularly neck or axilla), burn over a joint that may affect mobility/function of the joint
- Potentially complex burns—high-pressure steam burns; cold injury; burns due to ionizing radiation; if there is suspicion of non-accidental injury; the patient is very old, very young, or pregnant; circumferential burns; other associated problems, e.g. trauma, coexisting illness
- Electrical and chemical burns

❗ Consider referral to A&E for smoke inhalation even in the absence of significant burns.

If managing a burn in the community

- Check tetanus immunity, and give immunization ± prophylaxis as necessary—➔ p. 1089
- Cover the burn with a non-adherent dressing (e.g. Mepitel®). Apply a non-fibrous secondary absorbent dressing such as a dressing pad, and secure well with a lightweight conforming bandage or a tubular gauze bandage
- Refer if burns are not healed in 10–12d

Box 29.4 The 'rule of 9's'

❗ Ignore areas of erythema only

Palm	1%
Arm (all over)	9%
Leg (all over)	18%
Front	18%
Back	18%
Head (all over)	9%
Genitals	1%

The 'rule of nines' is not accurate for children <10y. For children and for small burns, estimate the extent of the burn by comparison with the area of the patient's hand. The area of the fingers and palm ≈1% total body surface area burn.

Secondary prevention of scalds and burns Children often sustain burns by pulling on the flex of boiling kettles or irons, pulling on saucepan handles, or climbing onto hot cookers; refer any child who has sustained accidental burns to the health visitor for follow-up.

Smoke inhalation Refer all patients who have potentially inhaled smoke for assessment—a seemingly well patient can deteriorate later. Smoke can cause:

- Thermal injury Airway problems occur due to thermal and chemical damage to the airways causing oedema—suspect if singed nasal hairs, a sore throat, or a hoarse voice
- Carbon monoxide poisoning May result in the classic cherry-red mucosa—but this may be absent
- Cyanide poisoning Commonly due to smouldering plastics and causes dizziness, headaches, and seizures

Sunburn Susceptibility depends on skin type.

- Tingling is followed 2–12h later by erythema. Redness is maximal at 24h and fades over 2–3d. Desquamation and pigmentation follow
- Severe sunburn may cause blistering, pain, and systemic upset. Treatment is symptomatic with calamine lotion prn (some advocate application of vinegar) and paracetamol for pain
- Rarely, dressings are required for blisters or, in severe cases, hospital admission for fluid management
- Predisposes to skin cancer and photoageing

The sun safety code Take care not to burn in the sun.

- Cover up with loose, cool clothing, a hat, and sunglasses
- If swimming outdoors or on the beach, dress in a UV protective sunsuit. When out of the water, add a T-shirt, sunglasses, and sun hat
- Seek shade during the hottest part of the day
- Apply sunscreen (≥ sun protection factor 25) on all sun-exposed parts of the body

Burns in special situations

Chemical burns

- Usually caused by strong acids or alkalis
- Wear gloves to remove contaminated clothing
- Irrigate with cold running water for ≥20min
- Do not attempt to neutralize the chemical—this can exacerbate injury by producing heat
- Refer all burns to A&E, unless the burn area is minimal and pain free

Electric shock

- Causes thermal tissue injury and direct injury due to the electric current passing through the tissue
- Skin burns may be seen at the entry and exit site of the current
- Muscle damage can be severe with minimal skin injury
- Cardiac damage may occur and rhabdomyolysis can result in renal failure
- Refer all patients for specialist management

Poisoning or overdose

On receiving a call for assistance

- Try to establish what has happened—substances involved, ongoing dangers, and state of the patient
- Arrange for the patient to be removed from any source of danger, e.g. contaminated clothing or inhaled gases. DO NOT put yourself or anyone else in danger attempting to do this. If necessary call the fire brigade, who have protective clothing and equipment, to help remove a patient from a dangerous environment
- If the patient is unconscious, immediately arrange for an ambulance to attend. Advise the caller to stay with the patient until help arrives.
- If the patient is conscious, advise to attend A&E immediately

Assessment of the unconscious patient Call for emergency ambulance assistance. Assess the need for basic life support:

- Airway patent?
- Breathing satisfactory?
- Circulation adequate?

⚠ Resuscitation (➔ p. 1040) takes priority over everything else.

Additionally

- If breathing is depressed and opioid overdose is a possibility, give naloxone 0.4–2mg IV every 2–3min to a maximum of 10mg (*child*—10 microgram/kg and then, if no response, 100 microgram/kg)
- Check capillary blood glucose—if low, give 50–250mL 10% glucose IV in 50mL aliquots

General examination

- BP
- Pulse
- Temperature
- Level of coma (➔ p. 1052)
- Pupil responses
- Evidence of IV drug abuse
- Obvious injury

❗ The coma may not be due to poisoning/overdose.

Turn into the recovery position Figure 29.4, ➔ p. 1043. Check no contraindications first, e.g. spinal injury.

Note down any information about the exposure

- Product name As much detail as possible—if unidentified tablets, see if any are left and send them to the hospital in their own container (if there is one) with the patient
- Time of the incident
- Duration of exposure/amount ingested
- Route of exposure Swallowed, inhaled, injected, etc.
- Whether intentional or accidental
- Take a general history from any attendant Medical history, current medication, substance abuse, alcohol, social circumstances

Assessment of the conscious patient

- Note down any information about the exposure as for the unconscious patient
- Record symptoms the patient is experiencing as a result of exposure

- Examine—pulse, BP, temperature (if necessary), level of consciousness or confusion, evidence of IV drug abuse, any injuries
- If non-accidental exposure assess suicidal intent (➔ p. 1100)
- Take a general history from the patient and/or any attendant—medical history, current medication, substance abuse, alcohol, social circumstances

⚠ Admit to A&E if:

- The patient's clinical condition warrants it e.g. unconsciousness, respiratory depression
- The exposure warrants admission for treatment or observation:
  - **Symptomatic poisoning**
  - **Agents with delayed action** Aspirin, iron, paracetamol, tricyclic antidepressants, co-phenotrope, paraquat, and modified-release preparations. Admit to hospital even if the patient seems well
  - **Other agents** Consult poisons information
- You judge there is serious suicidal intent (➔ p. 1100) or the patient has another mental health condition which warrants acute admission
- There is a lack of social support

Overdose and poisoning in children Peak incidence of accidental poisoning is at 2y—mainly household substances, prescribed or OTC drugs, or plants. Teenagers may take deliberate overdoses—especially of OTC medication, e.g. paracetamol.

⚠ Poisoning can be a form of non-accidental injury (➔ p. 902).

Deliberate self-harm (DSH) Deliberate, non-fatal act committed in the knowledge that it was potentially harmful and, in the case of drug overdose, that the amount taken was excessive. Self-harm is often aimed at changing a situation (e.g. to get a boyfriend back), communication of distress ('cry for help'), a sign of emotional distress, or may be a failed genuine suicide attempt.

Self-poisoning Accounts for 20% of admissions to general medical wards—the most frequent reason for admission for young ♀ patients. Paracetamol or aspirin are the most common drugs used.

Cutting Another common cause of self-harm, particularly among school-age children.

Management ➔ p. 1101

❗ People who have self-harmed should be treated with the same care, respect, and privacy as any other patient.

Poisons information

TOXBASE poisons database 🖱 www.toxbase.org (registration required)
UK National Poisons Information Service ☎ 0344 892 0111 (Ireland: (01) 809 2566)

Suicide and attempted suicide

Calls to patients who have deliberately self-harmed themselves, are threatening suicide, or if relatives are worried about suicide risk are common primary care emergencies.

- **Assessment** Figure 29.21
- **Management** Figure 29.22

If any self-harm Assess the situation and admit to A&E as needed

Ask about suicidal ideas and plans in a sensitive but probing way. It is a common misconception that asking about suicide can plant the idea into a patient's head and make suicide more likely. Evidence is to the contrary

Ask about present circumstances
What problems are making the patient feel this way?
Does he/she still feel like this?
Would the act of suicide be aimed to hurt someone in particular?
What kind of support does the patient have from friends, relatives, and formal services (e.g. community mental health services)?

Assess suicidal risk Ask patient and any relatives/friends present. *Risk factors*:

- ♂ > ♀
- Social isolation
- Depression
- Personality disorder
- Serious illness (e.g. cancer)
- Peak age 35–49y
- History of deliberate self-harm (100x ↑ risk)
- Alcohol or substance abuse
- Schizophrenia
- Contact with the criminal justice system
- Divorced > widowed > never married > married
- Certain professions: vets, pharmacists, farmers, doctors
- Admission or recent discharge from psychiatric hospital

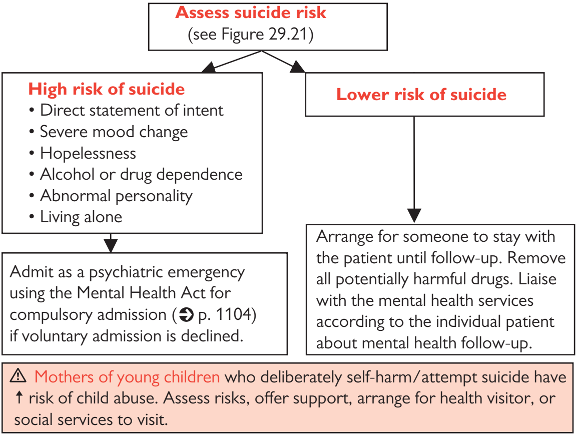
Assess mental health Features associated with ↑ suicide risk are:

- Depression
- Agitation
- Presence of suicidal ideation
- Hopelessness—good predictor of subsequent and immediate risk
- Early schizophrenia with retained insight—especially young patients who see their ambitions restricted
- Presence of delusions of control, poverty, and/or guilt

Useful questions for assessing suicidal ideas and plans

- Do you feel you have a future?
- Do you feel that life's not worth living?
- Do you ever feel completely hopeless?
- Do you ever feel you'd be better off dead and away from it all?
- Have you ever made any plans to end your life (if drug overdose—have you handled the tablets)?
- Have you ever made an attempt to take your own life? If so, was there a final act, e.g. suicide note?
- What prevents you doing it?
- Have you made any arrangements for your affairs after your death?

Figure 29.21 Assessment of patients who have deliberately self-harmed, or threatened or attempted suicide



Assess suicide risk (see Figure 29.21)

High risk of suicide
- Direct statement of intent
- Severe mood change
- Hopelessness
- Alcohol or drug dependence
- Abnormal personality
- Living alone

Admit as a psychiatric emergency using the Mental Health Act for compulsory admission (➔ p. 1104) if voluntary admission is declined.

Lower risk of suicide

Arrange for someone to stay with the patient until follow-up. Remove all potentially harmful drugs. Liaise with the mental health services according to the individual patient about mental health follow-up.

⚠ Mothers of young children who deliberately self-harm/attempt suicide have ↑ risk of child abuse. Assess risks, offer support, arrange for health visitor, or social services to visit.

Figure 29.22 Management of patients who have deliberately self-harmed, or threatened, or attempted suicide

Compulsory admission ➔ p. 1104

Suicide prevention In the UK, 1 in 5600 ♂ and 1 in 18,000 ♀ dies as a result of suicide. Suicide risk can be ↓ by:

- Early recognition, assessment, and treatment of those likely to attempt suicide—many visit their GP just weeks before suicide
- Planning follow-up care for those discharged from psychiatric hospitals
- ↓ availability and lethality of suicide methods, e.g. avoid tricyclic antidepressants and carefully monitor antidepressant repeat prescriptions

Further information

NICE (2004) Self-harm in over 8s: short-term management and prevention of recurrence. 🖱 www.nice.org.uk/guidance/cg16

NICE (2018) Preventing suicide in community and custodial settings. 🖱 www.nice.org.uk/guidance/ng105

RCGP/RCPsych Suicide mitigation in primary care factsheet. 🖱 www.connectingwithpeople.org/sites/default/files/SuicideMitigationInPrimaryCareFactsheet_0612.pdf

Information and support for patients and relatives

Harmless 🖱 www.harmless.org.uk
Samaritans ☎ 116 123 🖱 www.samaritans.org
Support after suicide partnership 🖱 http://supportaftersuicide.org.uk/

Disturbed behaviour

⚠ Look after your own safety

- If the patient is known to be violent, get back-up from the police before entering the situation
- Tell someone you are going in and when to expect an ‘exit’ call. Advise them to call for help if that call is not made
- Do not put yourself in a vulnerable situation—sit where there is a clear, unimpeded exit route
- Do not make the patient feel trapped
- Do not try to restrain the patient

Acute hyperventilation/panic attack ➔ p. 1081

Violent or agitated behaviour When a patient becomes very agitated or violent or starts to behave oddly, the GP may be called—by the patient, relatives or friends, or police attending the disturbance.

Causes of disturbed behaviour

- **Physical illness causing acute delirium** Infection (e.g. UTI, chest infection); hypoglycaemia (➔ p. 1082); hypoxia; head injury (➔ p. 1094); epilepsy (➔ p. 546)
- **Drugs** Alcohol (or alcohol withdrawal); prescribed drugs (e.g. steroid psychosis); illicit drugs (e.g. amphetamines)
- **Mental health problems** Schizophrenia; mania; anxiety/depression; dementia; personality disorder (e.g. attention-seeking; uncontrolled anger)

Assessment

- Before seeing the patient gather as much information as possible from notes, relatives—even neighbours
- Ask the patient and family for any history of drugs or alcohol excess
- Listen to the patient and talk calmly—choose your words carefully
- Try to look for organic causes—this can be difficult in the heat of the moment—physical examination except from a distance may be impossible. Do not put yourself at risk
- Suspect an organic cause where there are visual hallucinations

⚠ *Acute management*

- After assessing the problem, decide if hospitalization is required and whether this can be done on a voluntary or involuntary basis
  - If the patient is an immediate danger to self or others, admission is warranted
  - If the cause for the disturbed behaviour is unclear, admission for investigation is needed
- Discuss and explain your suggested management plan with the patient and any attendants
- Instigate management of treatable causes identified, e.g. admit if acute coronary syndrome or stroke is suspected; treat UTI or chest infection
- Consider sedation to cover the period before admission or to alleviate symptoms if admission is inappropriate

Suitable drugs to use for sedation

- Oral Diazepam 5–10mg po or lorazepam 1mg po/sublingually; chlorpromazine 25mg po (lower dose if elderly)
- Intramuscular Lorazepam 1.5–2.5mg; chlorpromazine 25mg; haloperidol 1–3mg

⚠ Avoid sedating patients with COPD, epilepsy, or if the patient has been taking illicit drugs, barbiturates, or alcohol.

Compulsory admission under the Mental Health Act ➔ p. 1104

❗ The Mental Health Act only allows for compulsory assessment and treatment of a patient's mental health problems—the patient may refuse consent for investigation and/or treatment of other health problems while 'sectioned'.

Acute dystonia Can occur soon after giving phenothiazines or butyrophenones, e.g. chlorpromazine, haloperidol. *Signs*:

- Torticollis
- Tongue protrusion
- Grimacing
- Opisthotonos (abnormal posturing with arching of the back)

Management Dystonia can be relieved with IM procyclidine 5–10mg (repeated as needed after 20min to a maximum dose of 20mg). Most GPs do not carry procyclidine, so refer urgently to A&E.

Delirium tremens (DTs) Major alcohol withdrawal symptoms usually occur 2–3d after an alcoholic has stopped drinking. *Features*:

- General Fever, tachycardia, ↑ BP, ↑ respiratory rate
- Psychiatric Visual/tactile hallucinations, acute delirium, apprehension
- Neurological Tremor, fits, fluctuating level of consciousness

⚠ *Action* DTs have 15% mortality—always admit as an emergency.

Compulsory admission and treatment of patients with mental illness

Most requiring inpatient care for mental disorder agree to hospital admission and become 'informal' patients. A minority (~5%) require compulsory admission and detention under the Mental Health Act of 1983 and 2007 and are termed 'sectioned' (in reference to the Section of the Mental Health Act under which they are detained)—Figure 29.23.

❗ The Mental Health Act applies in England and Wales only. In Northern Ireland, similar provisions apply under the Mental Health (Northern Ireland) Order 1986. Scotland—➔ p. 1105.

Procedure for 'sectioning' a patient

Applications can be made for

- Admission for assessment under Section 2 (➔ p. 1107)
- Admission for treatment under Section 3 (➔ p. 1107)
- Emergency admission under Section 4 (➔ p. 1107)
- Guardianship under Section 7 (➔ p. 1107)

Applications can be made by

- An approved mental health professional (AMHP), *or*
- The nearest relative of the person concerned. Nearest relative is defined in the Act as the 1st surviving person out of:
  1. Spouse (or cohabitee for >6mo)
  2. Oldest child (if >18y)
  3. Parent
  4. Oldest sibling (if >18y)
  5. Grandparent
  6. Grandchild (>18y)
  7. Uncle or aunt (>18y)
  8. Nephew or niece (>18y)
  9. Non-relative living with patient for ≥5y

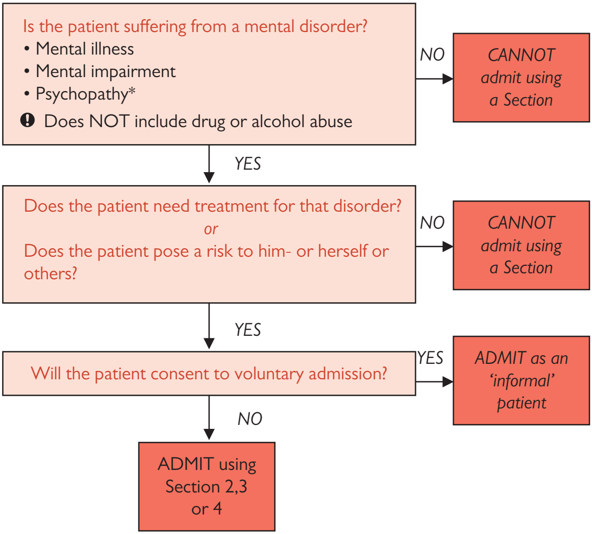
The applicant (AMHP or nearest relative) must have seen the patient <2wk (<24h in the case of Section 4) before the date of the application.

❗ The AMHP should be chosen rather than the nearest relative wherever possible, to avoid affecting family relationships.

Medical recommendations Applications must be based on 2 medical recommendations (except Section 4 which only needs one).

- Where 2 medical recommendations are required, the clinicians should not be from the same hospital or practice and one of the clinicians must be 'approved' under the Mental Health Act. One clinician, if practicable, should have prior knowledge of the patient (ideally a GP—but GPs are not obliged to attend outside the practice area). If neither clinician has prior knowledge of the patient, the applicant must state on the application why that was so. Clinicians may examine the patient together or separately, but there must be <6d between examinations
- Recommendations must be signed on or before the date of application; medical recommendation(s) and application must concur on at least one form of mental disorder

Relevant Sections of the Mental Health Act (2007) for GPs See Table 29.9, ➔ p. 1107.


Is the patient suffering from a mental disorder?
• Mental illness
• Mental impairment
• Psychopathy*
❗ Does NOT include drug or alcohol abuse
NO
CANNOT admit using a Section
YES
Does the patient need treatment for that disorder?
or
Does the patient pose a risk to him- or herself or others?
NO
CANNOT admit using a Section
YES
Will the patient consent to voluntary admission?
YES
ADMIT as an 'informal' patient
NO
ADMIT using Section 2,3 or 4

* Personality disorder characterized by inability to make loving relationships, antisocial behaviour, and lack of guilt

In practice 'sectioning' means calling in the duty social worker (or other social services approved mental health professional) and duty psychiatrist. It can be a time-consuming and frustrating business. Always try to obtain voluntary admission—it is better for you and the patient.

Keep a supply of forms you might need for sectioning—Forms 3, 7, and 10 (GP recommendation for Section 2, 4, and 3, respectively) and Form 5 (application for Section 4 for a 'nearest relative').

Deputizing doctors should always try to contact the patient's own GP.

Figure 29.23 Deciding whether a 'section' is needed

❗ The Mental Health Act only allows for compulsory assessment and treatment of a patient's mental health problems—the patient may refuse consent for investigation and/or treatment of other health problems while 'sectioned'.

Scotland The Mental Health Act (Care and Treatment) (Scotland) 2003 (and 2015 amendments) provides for compulsory admission under Part 5 for 72h. The application is made by a fully registered medical practitioner in consultation with a mental health officer, unless this is impracticable. In hospital, Part 6 (lasting 28d) can be applied and then, if necessary, Part 7 (Compulsory Treatment Order) for 6mo.

Mental Health Community Act (1995) Makes provision for supervision in the community after discharge from hospital for people with a mental disorder who are aged ≥16y and where:

- There could be serious risk of harm to the patient or others if the patient were not to receive further care services, *and*
- Supervision would help to ensure receipt of further care services

Application is made by the responsible medical officer (usually psychiatrist leading care) for 'after care under supervision' (ACUS) to the responsible health authority 6-monthly for the first year then yearly. ❗ If patients refuse treatment, they cannot be treated against their will but can be conveyed to a psychiatric day centre or hospital.

Deprivation of Liberty Safeguards (DoLS) Part of the Mental Capacity Act (MCA) (➔ p. 96). Deprivation of liberty is defined as circumstances 'where a person is under continuous control and supervision, is not free to leave and lacks capacity to consent to these arrangements'.

Applications for DoLS Made by a 'managing authority (usually care home or hospital) to a 'supervisory body' (usually local authority). In general, only applies when a patient does not object to care and applies to hospital or care home settings. Before application, it is important to:

- Assess the capacity of the individual to consent to arrangements for care/treatment in accordance with the MCA
- Consider whether the circumstances of the proposed accommodation and treatment amount (or are likely to amount) to a deprivation of liberty. Could the care plan be amended to avoid any potential deprivation of liberty?

Authorization is given If all 6 qualifying requirements are met:

1. Age ≥18y
2. The person has a qualifying mental health disorder or learning disability
3. The person currently lacks capacity to decide whether or not to be accommodated in the care home or hospital specified
4. It is proportionate, necessary, and in the best interests of the individual to be deprived of liberty
5. The individual is eligible for DoLS under the terms of the MCA
6. The person has not made a valid and applicable advance decision to refuse the care plan proposed, and no one else eligible to make decisions on the individual's behalf disagrees with the proposed plan.

❗ DoLS authorization does not authorize care or treatment, although this can be provided under the MCA in the individual's 'best interest'. DoLS authorizations should be notified to the Care Quality Commission.

Further information

Department of Health and Social Care (2015, updated 2017) Mental Health Act 1983: Code of practice. 🖱 www.gov.uk/government/publications/code-of-practice-mental-health-act-1983

Department of Health and Social Care (2015, updated 2018) Deprivation of liberty safeguards: resources. 🖱 www.gov.uk/government/publications/deprivation-of-liberty-safeguards-forms-and-guidance

Mental Welfare Commission for Scotland Mental Health Act. 🖱 www.mwcscot.org.uk/the-law/mental-health-act

Table 29.9 Sections of the Mental Health Act relevant to primary care

Section	Application
Section 2 *Admission for assessment* • Most commonly used section in the community • Admission for 28d for assessment • Not renewable after that time • Patients may appeal within 2wk of detention via the Mental Health Tribunal	• Application must be made by the nearest relative or an AMHP on the recommendation of 2 doctors—1 approved and the other who has prior knowledge of the patient • If application is made by the AMHP, the nearest relative should be informed before application or as soon as possible afterwards • Application is valid for 14d
Section 3 *Admission for treatment* • Admission for treatment for ≤6mo • The exact mental disorder must be stated • Detention is renewable for a further 6mo and then annually	• Application must be made by the nearest relative or an AMHP on the recommendation of 2 doctors—1 approved and the other who has prior knowledge of the patient • Application is valid for 14d
Section 4 *Emergency admission for assessment* • Used where admission is urgent and compliance with Section 2 would cause undesirable delay • Admission to hospital for 72h only • Not renewable • Usually converted to a Section 2 on arrival at hospital	• Application must be made by the nearest relative or an AMHP; if application is made by the AMHP, the nearest relative should be informed before application or as soon as possible afterwards • Medical recommendation is from either an approved clinician (not necessarily a doctor) or a doctor with prior knowledge of the patient • Application is only valid for 24h
Section 7 *Guardianship*. A guardian has power to: • Require a person to live at a particular place • Require a person to go to specific places at specific times for medical treatment, work, education, or training • Require a doctor, AMHP, or other specified person be given access to the person under Guardianship ❗ Guardians can insist a person sees a doctor but cannot force treatment	• Application must be made by the nearest relative or an AMHP on the recommendation of 2 clinicians—1 approved and the other who has prior knowledge of the patient • Application is valid for 14d
Section 115 Allows an approved mental health professional to enter and inspect any premises (except hospital) in which a person with a mental disorder is living if there is reasonable cause to believe that person is not under proper care Application through a magistrate is needed	
Section 135 Gives right of entry of a police officer who believes a person with a mental disorder is being ill-treated or suffering from self-neglect to enter premises and remove that person to a place of safety. The police officer must be accompanied by an approved mental health professional and approved clinician unless the person is already 'sectioned' and absent without leave. Requires application to a magistrate	

Miscellaneous and environmental emergencies

Acute stroke and intracranial bleeding ➲ p. 534

Immediate management of stroke If stroke is suspected, admit directly to hospital by emergency ambulance. Thrombolysis early after ischaemic stroke results in better outcomes, so do not delay referral until the patient is seen. Treatment of stroke in a stroke unit ↓ mortality and morbidity[G].

⚠ Do not give aspirin prior to admission.

Acute limb ischaemia *Causes*:
- Acute thrombotic occlusion of pre-existing stenotic segment (60%)
- Embolus (30%)
- Trauma, e.g. compartment syndrome or traumatic vessel damage

Presentation The 6 'P's:
- Pain
- Pallor
- Paraesthesiae
- Pulselessness
- Paralysis
- Perishing cold

Action Admit acutely under the care of a vascular surgeon. Treatment can be surgical (e.g. embolectomy) or medical (e.g. thrombolysis).

Wound dehiscence Breakdown of a surgical wound—usually abdominal. May be partial or complete. *Risk factors*:
- Malnutrition
- Obesity
- ↑ intra-abdominal pressure, e.g. from coughing
- Wound infection
- Haematoma formation
- Ascites draining through a wound

Partial breakdown Skin remains intact but muscle layers break down resulting in incisional hernia. Typically the patient feels something 'give' ± sudden ↑ in pain and pink fluid discharge. Refer for urgent reassessment by the operating surgeon.

Complete dehiscence Wound breaks down entirely. The patient becomes shocked and distressed. Lie flat; give strong opioid analgesia; cover the wound with a sterile pack soaked in saline or cling film if more readily available; admit as a 'blue light' emergency.

Drowning Most common in drunk adults and children poorly supervised around water. Children can drown in a few centimetres of water.

Action
- Call for help
- Start basic life support (Airway, Breathing, Circulation)—➲ p. 1040 (adults); ➲ p. 1044 (children)

⚠ Attempted resuscitation of a seemingly dead child is worthwhile as cooling ↓ metabolic rate and recovery can occur even after prolonged immersion in cold water.

Prevention Drowning is the third most common cause of accidental death among the under 16s. More than half of those who drown can swim. Most people drown in rivers (25%) or the sea (17%) but, for children aged <4y, garden ponds are the most common place of drowning. For adults, alcohol is a contributory factor in 25–50% cases. The best way to ↓ drowning is prevention—spot the dangers; take safety advice; do not go near water alone; learn how to help others.

Hypothermia Defined as a core temperature of <35°C. *Causes*:

- Immobility
- Hypothyroidism
- DM
- Alcohol
- Not feeling the cold, e.g. neuropathy, confusion, dementia
- Inadequate heat in the home, e.g. poor housing, poverty, and fear of high fuel bill
- Inadequate protection from the cold, e.g. unsuitable clothing while doing outdoor sports
- Drugs—antipsychotics, antidepressants, barbiturates, tranquillizers—may lower the level of consciousness and ↓ ability to shiver
- Falls—may remain still and cold on the floor until discovered
- Unconsciousness, e.g. overdose, stroke

Presentation Skin pale and cold to touch; puffy face; listlessness, drowsiness, and/or confusion. *When severe:* ↓ breathing—slow and shallow; ↓ pulse volume—faint and irregular; stiff muscles; loss of consciousness.

Investigation Often not possible in the acute situation in the community. Go on history and signs. If equipment is available:

- Rectal temperature on low-reading thermometer <35°C
- ECG—'J' wave on the end of the QRS complex

Action

- Remove the patient from the cold environment if possible, or wrap in blankets/coats—including head. ❗ Do not use direct heat (e.g. hot water bottles), as this can cause rapid fluid shifts and potentially fatal pulmonary oedema
- Transfer to hospital as an emergency
- Consider the cause of the incident; liaise with the hospital, wider primary healthcare team, and social services to prevent recurrence

Heat stroke and heat exhaustion Exercising in excessive heat leads to dehydration, salt depletion, and metabolite accumulation.

- Signs Headache, nausea, confusion, incoordination, cramps, weakness, dizziness, malaise
- Treatment Rest, fluid, and salt replacement. Admit for IV fluids and supportive measures in severe cases

Sunburn ➜ p. 1097

Acute altitude sickness Altitude sickness is a potentially fatal complication of rapidly climbing to altitudes >2500m (8000 feet). 2 main forms: pulmonary oedema and cerebral oedema. Presents with fatigue, headache, dizziness, nausea/loss of appetite, breathlessness, palpitations, and/or insomnia. Treatment is with oxygen therapy and descent to a lower altitude. Prevent by gradual ascent. 💣 Use of prophylactic acetazolamide is controversial.

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Reference intervals

❗ These are guides only—different laboratories may use different units and ranges. Pregnant women and children also have different normal ranges—consult your local laboratory for further information.

Biochemistry		*Reference interval*
Sodium	P	135–145mmol/L
Potassium	P	3.5–5.0mmol/L
Bicarbonate	P	24–31mmol/L
Creatinine	P	60–125 micromol/L
Estimated glomerular filtration rate (eGFR) ➔ p. 410	P	>90mL/min is normal
Calcium (total corrected)	P	2.15–2.55mmol/L
Urea	P	3.0–6.5mmol/L
Osmolality	P	280–295mosmol/kg
Creatinine kinase (CK)	P	♂ 25–195IU/L; ♀ 25–170IU/L
Troponin I	P	<0.15 micrograms/L
—*Acute coronary syndrome*		*>1.5 micrograms/L*
Phosphate (inorganic)	P	0.7–1.5mmol/L
Amylase	P	70–330U/dL
Protein (total)	P	63–80g/L
Albumin	P	32–47g/L
Bilirubin	P	0–17mmol/L
Alkaline phosphatase	P	100–300U/L
Aspartate transaminase (AST)	P	5–42IU/L
Alanine aminotransferase (ALT)	P	5–42IU/L
Gamma-glutamyl transpeptidase (GGT)	P	♂ 10–46IU/L; ♀ 6–29IU/L
Uric acid (urate)	P	♂ 0.15–0.45mmol/L; ♀ 0.12–0.36mmol/L
Glucose (fasting) ➔ p. 317	P	4.0–6.0mmol/L
HbA1c ➔ p. 317	P	<48mmol/mol
—*Pre-diabetes*		*44–48mmol/mol*
Total cholesterol ➔ p. 222	P	Ideally <5.0mmol/L
—*LDL cholesterol*		*Ideally <3.0mmol/L (<2.0 if high risk)*
—*HDL cholesterol*		*Ideally >1.0mmol/L*
—*Total cholesterol:HDL*		*Ideally <4*
Triglyceride ➔ p. 222	P	<2.1mmol/L
Vitamin B_{12}	P	180–1000ng/L
Folate	S/P	3–17 micrograms/L
Iron	S	♂ 14–33mmol/L; ♀ 11–28mmol/L
Ferritin	P	10–120 micrograms/L pre-menopausal ♀ 14–200 micrograms/L post-menopausal ♀ and ♂
Prolactin	P	♂ <450U/L; ♀ <600U/L
Prostate-specific antigen (PSA) ➔ p. 433	P	<50y 0–2.5 micrograms/L 50–59y 0–3.5 micrograms/L 60–69y 0–4.5 micrograms/L >70y 0–6.5 micrograms/L
Thyroxine (free T_4)	P	8–22pmol/L
TSH	P	0.35–5.5mLU/L

P = plasma (e.g. heparin bottle); S = serum (clotted—no anticoagulant).

Sex hormones		Reference interval	
		FSH (IU/L)	LH (IU/L)
Adult ♀	*Follicular phase*	1–9	1–12
	Ovulatory phase	6–26	16–104
	Luteal phase	1–9	1–12
	Post-menopausal	30–118	16–66
Adult ♂		1–7	1–8

Haematology	*Reference interval*
Haemoglobin	♂ 13.0–17.0g/L; ♀ 12.0–15.0g/L
Anaemia ➔ p. 638	
Erythrocytosis ➔ p. 655	
Red cell count (RCC) or erythrocytes	♂ 4.5–5.5 × 10^{12}/L; ♀ 3.8–4.8 × 10^{12}/L
Packed cell volume (PCV) or haematocrit	♂ 0.40–0.50 L/L; ♀ 0.36–0.46 L/L
Mean cell volume (MCV) ➔ p. 636	80–100fL
Mean cell haemoglobin (MCHC)	32.0–36.0g/dL
White cell count (WCC) ➔ p. 637	4.0–11.0 × 10^9/L
Neutrophils	*2.0–7.5 × 10^9/L (40–75% WCC)*
Lymphocytes	*1.5–4.0 × 10^9/L (20–45% WCC)*
Eosinophils	*0.04–0.50 × 10^9/L (1–6% WCC)*
Basophils	*0.02–0.10 × 10^9/L (0–1% WCC)*
Monocytes	*0.2–1.0 × 10^9/L (2–10% WCC)*
Platelet count ➔ p. 636	150–400 × 10^9/L
Reticulocyte count * Only use % if RCC is normal	♂ 25–135 × 10^9/L (0.5–2.5%* RCC) ♀ 20–120 × 10^9/L (0.5–2.5%* RCC)
Erythrocyte sedimentation rate (ESR) ➔ p. 636	<50y ♂ 10mm, ♀ 19mm 51–60y ♂ 12mm, ♀ 19mm 61–70y ♂ 4mm, ♀ 20mm >70y ♂ 30mm, ♀ 35mm
International normalized ratio (INR)	Therapeutic ranges—➔ p. 648

NEWS2 Score for adults

Physiological parameter	Score						
	3	2	1	0	1	2	3
Respiration rate (per minute)	≤8		9–11	12–20		21–24	≥25
SpO_2 Scale 1 (%)	≤91	92–93	94–95	≥96			
SpO_2 Scale 2 (%)	≤83	84–85	86–87	88–92 ≥93 on air	93–94 on oxygen	95–96 on oxygen	≥97 on oxygen
Air or oxygen?		Oxygen		Air			
Systolic blood pressure (mmHg)	≤90	91–100	101–110	111–219			≥220
Pulse (per minute)	≤40		41–50	51–90	91–110	111–130	≥131
Consciousness				Alert			CVPU
Temperature (°C)	≤35.0		35.1–36.0	36.1–38.0	38.1–39.0	≥39.1	

Reproduced from: Royal College of Physicians. *National Early Warning Score (NEWS) 2: Standardising the assessment of acute-illness severity in the NHS*. Updated report of a working party. London: RCP, 2017

Anaphylaxis algorithm

Anaphylactic reaction?

↓

Check/record:
A—airway—lip/throat swelling
B—breathing—respiratory rate, peripheral oxygen saturation, wheeze/stridor
C—circulation—BP, pulse rate, cyanosis
D—disability e.g. distress, confusion, coma
E—exposure e.g. skin changes, angio-oedema

↓

Call for help and an emergency ambulance
If possible, lie flat

↓

Give IM1:1000 adrenaline using a ≥25mm needle—dose:
Adult and child aged >12y: 0.5mL (500 micrograms)
Child aged 6–12y: 0.3mL (300 micrograms)
Child aged <6y: 0.15mL (150 micrograms)
Repeat after 5min if no improvement

When skills and equipment are available:

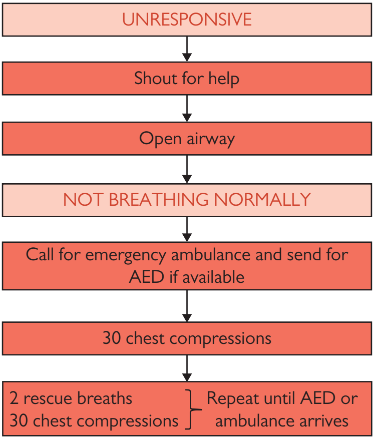
Monitor:
BP, ECG, pulse oximetry

Consider giving:

IV fluid challenge:	**Chlorphenamine** (IM or slow IV):	**Hydrocortisone** (IM or slow IV):
Adult: 500mL –1L *Child*: crystalloid 20mL/kg STOP any IV colloid	*Adult or child aged >12y*: 10mg *Child aged 6–12y*: 5mg *Child aged 6mo–6y*: 2.5mg *Child aged <6mo*: 250 micrograms/kg	*Adult or child aged >12y*: 200mg *Child aged 6–12y*: 100mg *Child aged 6mo–6y*: 50mg *Child aged <6mo*: 25mg

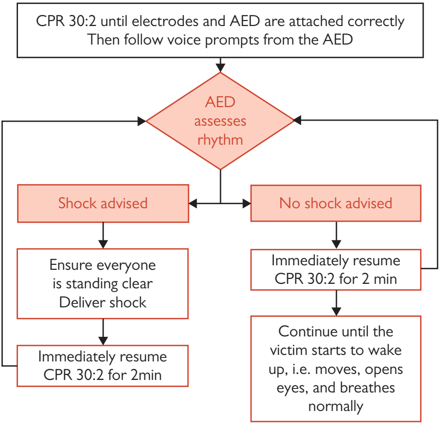
Source: data from Emergency treatment of anaphylactic reactions: Guidelines for healthcare providers, https:// www.resus.org.uk/ anaphylaxis/ emergency-treatment-of-anaphylactic-reactions/

Adult basic life support (ABLS) algorithm


UNRESPONSIVE
Shout for help
Open airway
NOT BREATHING NORMALLY
Call for emergency ambulance and send for AED if available
30 chest compressions
2 rescue breaths
30 chest compressions
Repeat until AED or ambulance arrives

❗ Give 2 rescue breaths for every 15 chest compressions for children

Automated external defibrillator algorithm


CPR 30:2 until electrodes and AED are attached correctly
Then follow voice prompts from the AED
AED assesses rhythm
Shock advised
No shock advised
Ensure everyone is standing clear
Deliver shock
Immediately resume CPR 30:2 for 2min
Immediately resume CPR 30:2 for 2 min
Continue until the victim starts to wake up, i.e. moves, opens eyes, and breathes normally

Paediatric life support algorithm ➔ p. 1045

Newborn life support algorithm ➔ p. 1087

Source: data from Adult basic life support and automated external defibrillation, https://www.resus.org.uk/resuscitation-guidelines/adult-basic-life-support-and-automated-external-defibrillation/